

RADIOLOGY

A MONTHLY JOURNAL DEVOTED TO CLINICAL RADIOLOGY AND ALLIED SCIENCES

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No. 5

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RADIOLOGY

A MONTHLY PUBLICATION DEVOTED TO CLINICAL RADIOLOGY AND ALLIED SCIENCES

PUBLISHED BY THE RADIOLOGICAL SOCIETY OF NORTH AMERICA

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Hip Lesions of Infants and Children Seen at the Newington Home and Hospital for Crippled Children¹

GILBERT W. HEUBLEIN, M.D., LOUIS BERNSTEIN, M.D., and B. J. HUBENET, M.D.

Hartford, Conn.

IN HIS ADDRESS at the Hartford Medical Society in June 1909, Arthur T. Legg (43), then Junior Assistant Surgeon of the Children's Hospital in Boston, made the following pertinent remarks regarding the well known affection of the hip which now bears his name: "We are at times painfully aware of the fact that there are many symptoms which we readily recognize in our clinical observations to which we can assign no cause, and it is also an undoubted fact that there are many conditions even which exist today of which we are ignorant, simply from our neglect to observe, or from faulty observation, or again, from faulty deduction even from good observation."

We are convinced of the fundamental importance of the attitude to which Legg gave expression in these words, particularly as it affects roentgen diagnosis. Even a superficial perusal of the literature convinces one of the complexity and breadth of the subject of hip disease. Therefore, much can be learned through accurate observation and deduction. Efficiency in the field of roentgen diagnosis can be attained only through thorough familiarity with all phases of roentgen technic and avoidance of abstract film reading without proper clinical control.

EMBRYOLOGICAL AND ANATOMICAL CONSIDERATIONS

According to Sir Arthur Keith (37), in the eighth week of intra-uterine existence (20 mm. fetus) the three principal components of the innominate bone meet to form the Y-shaped acetabular suture. In the ninth week, outgrowth of cartilage from the primitive ischium, ilium, and pubis results in the formation of the cotyloid cavity. A synovial lining then appears. In certain cases in the reptilian stage (second month) the acetabular cup fails to develop, and congenital dislocation of the femoral head follows. Similarly, Keith states that cleft palate and imperforate anus may result from arrest of development in this stage. Apparently development of congenital dislocation of the hip has a definite relation to female sexual characters, since the majority of cases (90 per cent) are encountered in female patients.

The importance of a knowledge of normal limb bud rotation as an aid to understanding certain hip joint deformities has been recently emphasized by Badgley (1). To simulate the position of the fetal limb buds, the adult should be placed prone with the limbs projecting fin-like at right angles from the trunk; the knees would then face

¹ Presented at the Thirty-third Annual Meeting of the Radiological Society of North America, Boston, Mass., Nov. 30-Dec. 5, 1947.

dorsad with the great toes in front (Keith), owing to external rotation. By comparison, the arms would be rotated internally so that the elbows face dorsad. As growth occurs, both limb buds rotate in opposite directions about their long axes. Thus, the elbow ultimately points posteriorly after external rotation and the knee ventrally after internal rotation; the palms thus face ventrad in the anatomical position and the soles dorsad. Finally, as the limb bud rotates from flexion to extension, reciprocal forces come into play at the hip joint. The socket is deepened posteriorly, and anteversion of the femoral head and neck simultaneously occurs. The caudal angle of the acetabulum increases and the ventral angle diminishes. The degree of anteversion varies from 0° in the first two months to 35° at birth, thereafter gradually diminishing to 15° or less. By way of clarification of the above, Badgley, in a personal communication (2) states: "The caudal angle of the acetabulum runs from the ischial portion of the sacrum. The ventral angle is made up primarily of the pubic bone. There is an increase in growth in the embryo of this posterior portion so that the acetabulum is altering its shape constantly, or rather its direction, lateral to an anterolateral direction because of the increased growth of the caudal portion. There is a lessened growth of the pubis. This, therefore, tends to place the acetabulum in a more oblique direction, facing more frontally than it did in the embryo. This is in keeping with the developing anteversion from 0 to 35° in the embryo. I think this is possibly a factor in the development of the anteversion. It certainly is a factor in the tendency toward its location. Comparative anatomy shows that this inclination forward is far greater in humans than in animals."

From the foregoing, it is clear that months of molding are ordinarily required before the normal acetabulum is completely formed. As demonstrated by Badgley, disturbances preventing normal rotation of the limb bud may result in various types of congenital dislocation.

ANATOMY OF THE HIP JOINT

The acetabulum is triradiate in shape, one-fifth being formed by the pubic portion, the other four-fifths, in equal proportions, by the ischial and iliac components. According to Köhler, these ossific centers appear between the fifth and ninth fetal months. Synostosis is not complete until the fourteenth to seventeenth year. This cartilaginous synchondrosis is readily distinguished in anteroposterior roentgenograms, affording a well known landmark for localization of the "Y" line and the "Y coordinate" (Ponseti, 59). In anteroposterior films the broad band-like defect of the synostosis becomes largely obliterated by the fifth year, although complete fusion does not occur until later. According to Grant (29), the fibers of the capsule, which were originally parallel, present a spiral pattern consequent to extension of the joint which occurs when man assumes the erect posture. Internally the capsule presents a mid-line constriction, produced by a belt of deep fibers encircling the fibrous joint capsule. This hour-glass constriction is known as the *orbicular zone* and is readily demonstrated in normal arthrograms made with 30 per cent ténébryl or similar contrast substances. It must not be confused with the hour-glass defect in congenital dislocation which, according to Leveuf (45), is produced by an hypertrophied and depressed cartilaginous limbus. It is noteworthy that many of the important non-opaque soft-tissue structures of the hip joint can be demonstrated by means of opaque arthrography. Severin (69) in a recent excellent publication has shown that the following structures can be identified: (a) the fibrocartilaginous limbus; (b) the orbicular zone; (c) the rounded impression of the transverse ligament; (d) the medially placed ventral and dorsal recesses of the acetabular fossae, small finger-like projections of the synovial capsule which enclose the base of the ligamentum teres and therefore indicate its site of origin. Severin stresses particularly the importance of identifying the free border of the cartilaginous acetabulum.

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This provides a means of distinguishing the normal joint from conditions which exist in a partial or complete luxation. He states: "If the joint is to be styled the normal, the border of the cartilaginous acetabulum should reach down laterally to the horizontal line through the Y-shaped cartilage or in its immediate vicinity. If this border is displaced upwards, a subluxation has developed, and when the caput has come away completely from the acetabulum a complete dislocation results."

The ligamentum teres, actually a flat structure, was first recognized in 130 B.C. (15); it spans the medial and inferior portion of the hip joint. Laterally it is attached to the superior portion of the fovea and inferomesially to the transverse ligament and adjacent margins of the acetabulum. Chandler and Kreuzsch (15) have pointed out that vessels carrying a significant blood supply either traverse the substance of the ligament or pass along its dorsal surface to anastomose ultimately with vessels of the femoral head. These vessels in the spongiosa make the femoral head relatively independent of the capsular arteries (Quist-Hansson, 62). Wolcott (77) reaches a similar conclusion: that the ligamentum teres, carrying as it does a consequential blood supply, plays an important role in preventing aseptic necrosis of the femoral head even in adults up to eighty years of age or older. Under twenty, anastomosis between the vessels of the ligamentum teres and capsular arteries has not developed. Therefore, ischemic necrosis following trauma is a relatively frequent occurrence in individuals under eighteen to twenty. Although the ligamentum teres is not demonstrated by direct roentgen study, one can suspect a complete tear of this structure after forcible extrusion of the femoral head from its socket, as well as in cases of reduced dislocation with lineal fracture of the posterior acetabular margin, or in central dislocation of the head. Following trauma, the region of the fovea should be carefully inspected for evidence of localized demin-

eralization and patchy areas of increased density, indicating impaired nutrition secondary to injury of the ligamentous arteries. Various observers (15, 77) have pointed out that pinning or drilling in the neighborhood of the fovea may compromise the blood supply of the femoral head. It should be recalled that in the hip, as elsewhere, three to four months must elapse before bone intensification changes indicating impaired nutrition can be detected by roentgen study.

The foregoing is an attempt to describe some of the important features relating to blood flow along the ligamentum teres vessels from the proximal aspect of the hip joint. The circulation from the distal aspect of the joint is derived from nutrient branches of the anterior and posterior circumflex femoral arteries which pass along the capsule and finally anastomose with the vessels of the spongiosa (62). Gall and Bennett (25) have shown that in the period of active growth, between five and twelve years, the nutrition of the capital epiphysis is particularly vulnerable. Circulation in the ligamentum teres is said to diminish considerably between the ages of two and five. They make the following important statement: "During the succeeding period of growth the capital epiphyseal cartilage remaining active and unfused serves as a barrier to the rich vascular bed of the marrow cavity and neck. The major source of nutrient supply consists therefore of small vessels which arise near the insertion of the capsule, are reflected subperiosteally along the neck of the femur, and enter the head somewhere in the vicinity of the margin of the articular cartilage. It has been suggested that the fineness of these vessels and their location cause them to be particularly susceptible to trauma." The above described fragility of vessels plus diminished blood supply through the ligamentum teres may serve as an anatomical explanation for the age incidence of osteochondropathy of the hip (Legg-Perthes disease). The roentgenologist particularly interested in the blood supply of the hip is referred to the excellent illustrations and

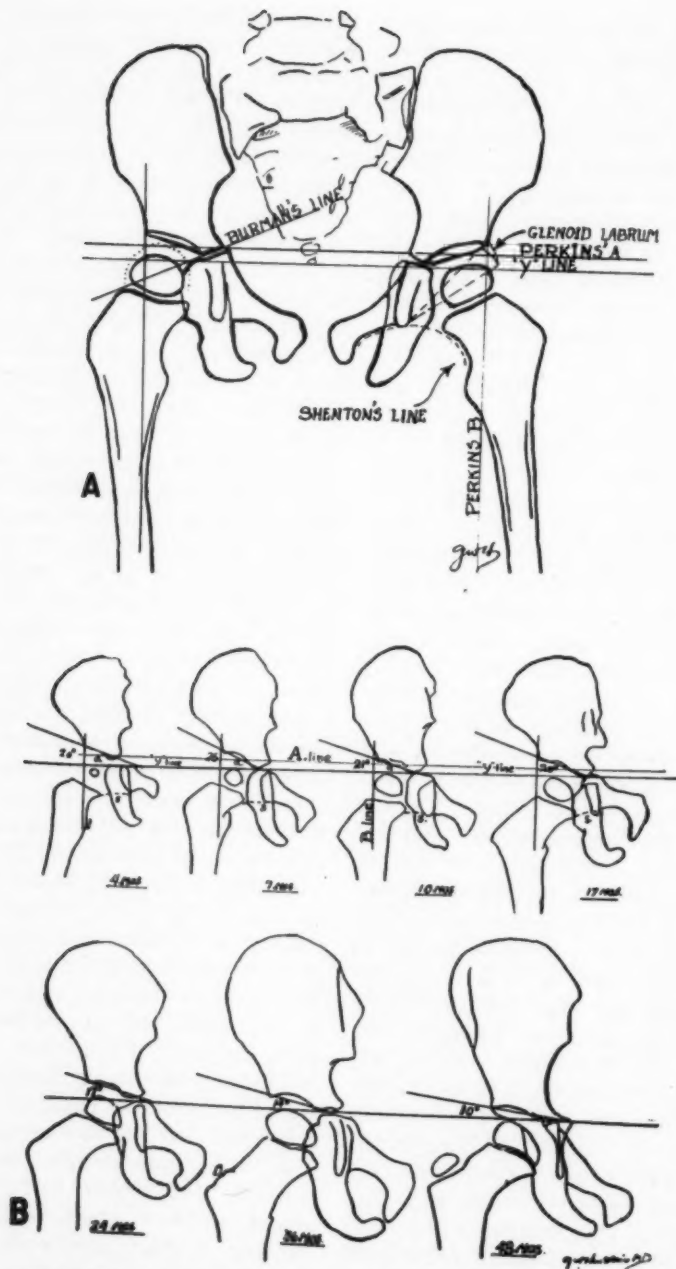


Fig. 1. The upper illustration (A) shows various methods used to localize the bony femoral epiphysis. It will be observed that the A and B lines of Perkins enclose the ossified portion of the femoral head. The Y-line lies on the ischial side of the triradiate cartilage. Burman's line is a continuation of the roof of the "Y-cartilage" and should transect the femoral head as shown. Note the smooth arc formed by Shenton's line.

Below are drawings (B) of normal hip joints between birth and 48 months. The angle of acclivity (a) decreases quite rapidly, from 20° at birth to 10° at 48 months. "S" indicates the position of the pubocoxofemoral line of Shenton. It will be observed that the bony nucleus of the femoral head lies in the inferolateral quadrant of the A and B lines.

text in "Lectures on Regional Orthopedic Surgery and Fundamental Orthopedic Problems" (21).

ADDITIONAL CONSIDERATIONS WITH SPECIAL REFERENCE TO ROENTGEN ANATOMY

The femoral capital epiphysis commonly appears during the end of the second half of the first year of life, although it may rarely be encountered in the newborn. According to Wiberg (76), the femoral head comprises two-thirds of a sphere. It is "practically spherical in shape." The apparent bony acetabulum in children roughly forms a sector of a circle, the angle of which increases with age. In a limited number of measurements it has been found that the angle of this sector subtends an arc of 60° at seven months and 123° at sixty months. In anteroposterior roentgenograms of the normal hip in children the osseous acetabulum encompasses one-third to one-half of the bony nucleus of the femoral head. If the tip of the cartilaginous labrum is projected into the vicinity of the "Y" line, then one-half to two-thirds of the ossified capital epiphysis will be found to be included in most instances (Fig. 1, A). Samuel (67) indicates that in the normal adult hip one-half to five-eighths of the femoral head will lie medial to a line connecting the upper and lower margins of the bony acetabulum. Slight anatomical variations are easily overlooked. Early recognition of minor variations of hip joint incongruence, such as a too deep or shallow socket, may enable one to anticipate and partially forestall the development of osteoarthritis in later life. The shallow socket can be recognized by the fact that the line of the bony roof of the acetabulum is more widely extended than usual. The radius of its arc becomes increased laterally, and the contour of the socket suggests a half lemon, instead of a half orange, a comparison first suggested by Calot (12).

Köhler's "tear-drop" or "pear-figure" (40) is an important landmark (Fig. 1). The outer line usually corresponds to the acetabular floor, while the inner line rep-

resents a tangential projection of the inner bony wall of the pelvis. Interruption of these lines is of pathologic significance. Burman and Clark (9) refer to this figure as the outer and inner accented lines. Jansen (36) says that a V-shaped pear-figure occurs quite frequently in coxa vara deformities. His explanation is that pressure transmitted through the femoral head *in utero*, with the thigh abducted, produces a twisted socket. This latter statement requires further study for confirmation. The wide tear-drop is, on the other hand, frequently encountered in shallow or dysplastic acetabula and is associated with a broad "Y" cartilage as seen in the anteroposterior projection, often with an irregular hyperplastic acetabular floor. Another method of estimating the proper position of the femoral head has been described by Burman and Clark (9). They were the first to call attention to the importance of the lateral prolongation of the "short iliac arm." This line, tangential to the roof of the "Y" cartilage in anteroposterior roentgenograms, runs downwards and outwards, its prolongation transecting or passing lateral to and above the capital epiphysis. Displacement of the bony nucleus of the femoral head above this line is a distinctly abnormal finding (Fig. 1, A).

The importance of the acetabular index of normal and dysplastic hips is worthy of comment. This angle shown at *a* in Figure 1, B, reflects the acclivity of the hip socket. According to Kleinberg and Lieberman (39), the degree of inclination is an index of "the resistance which the iliac portion of the acetabulum would present to luxation of the head of the femur." Actually this line drawn from the outer margin of the apparent acetabular roof, downwards and inwards to a point where the iliac bone joins the "Y" line, does not correspond to the true limits of the acetabulum, which is cartilaginous and, therefore, not seen. From the foregoing, a certain amount of skepticism regarding the importance of this measurement would seem justifiable. However, observation of 300 normal hips studied on films picked at random from

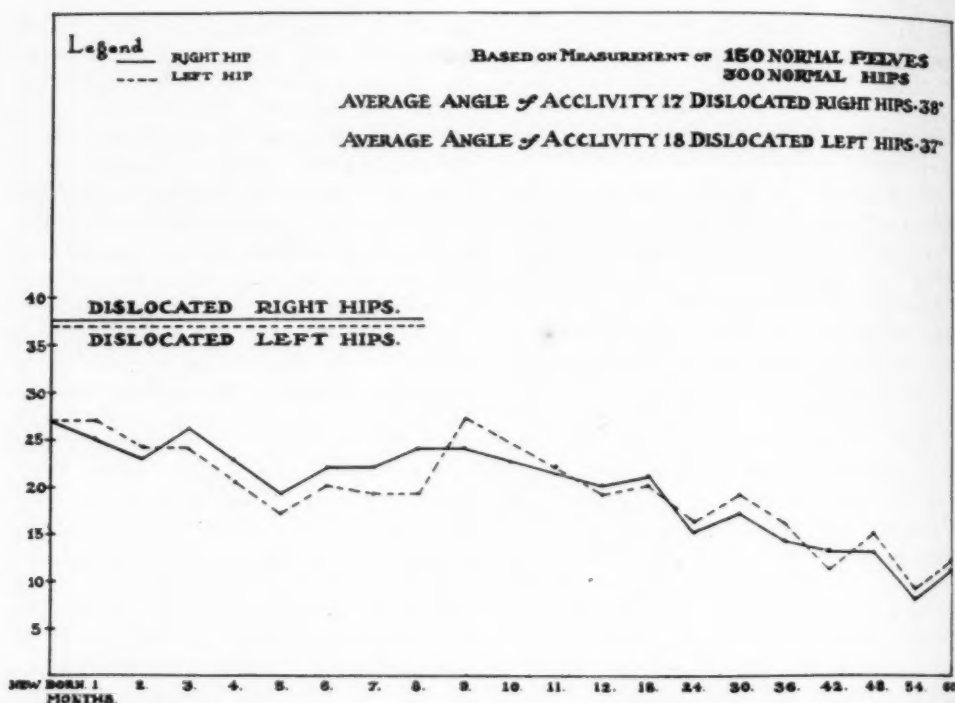


Fig. 2. Graph showing the reduction in the angle of acclivity between birth and 60 months. At 12 months the slope of the acetabulum has decreased to 19–20°, whereas at 60 months it is only 9–10°. By comparison, the angle of acclivity of 35 dislocated hips was found to be on the order of 37–38°.

our stacks indicates that, depending upon the age of the patient, the acetabular index is relatively constant and is of aid in evaluating the normal acetabulum in infants and children. Our findings (Fig. 2) are practically identical with those described by Kleinberg in the first months of life, and our measurements have been extended to include children up to six years of age.

In infants of nine days or under, we have found the average acclivity of 67 hips to be 27° with a range of 10 to 44°. The average acetabular index diminishes with age, so that at twelve months it is found to be 19 or 20°, and at sixty months has diminished to 11 or 12° (Fig. 2).

In view of the measurements referred to above (60° at seven months, 123° at sixty months), it is understandable that in young children, due to incomplete ossification of the acetabular roof, the configuration of the socket may simulate the picture of a

dysplastic acetabulum in older age groups (demi-citron shape). Unless the deformity is very definite, therefore, in subjects under four years, care should be taken to avoid a false positive diagnosis.

The normal radiographic appearance of the pelvis and hips in very young children is not generally appreciated. Perkins (54), in 1928, was among the first to call attention to the importance of localizing the position of the bony capital epiphysis in relation to a horizontal line transecting the "Y" cartilages (Perkins' A line) and vertical lines dropped from the anterior inferior iliac spines at right angles to it (Perkins' B lines) (see Fig. 1, A). Hart (31) recommends drawing the "Y" line through the gap presented by the "Y" cartilages but tangent to the ischial side of the joint, that is, slightly below the level originally advocated by Perkins. Whatever means of localization is employed, it

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will be found that normally the bony nucleus of the femoral head lies below the "Y" line and medial to Perkins' B line. As shown in Figure 1, A, the nucleus of the head lies in the inferomesial quadrant of the arbitrarily constructed cross (Burman and Clark). It is noteworthy that the superior and lateral margins of the capital epiphysis are tangent to both lines in a surprisingly high percentage of cases.

From a study of dissections of stillborn infants made by one of us (B. J. H.) at the Yale School of Medicine, the following facts have appeared:

nucleus in the center of a large radiolucent femoral head.

CASE HISTORIES

Early diagnosis of hip disease in infancy and childhood constitutes a challenge to the surgeon and radiologist on two scores. The anatomical components of the hip in the newborn, as shown above, are largely cartilaginous (Fig. 3), rendering many important structures invisible to direct inspection. Therefore, painstaking examination of anatomical relationships is required, especially of the proximal end of

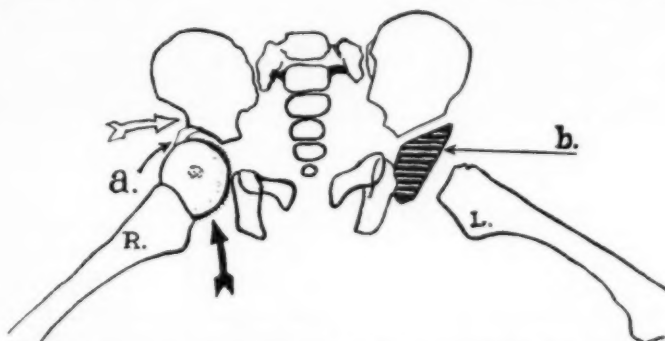


Fig. 3. Composite drawing showing the appearance of the hips at birth. The left hip socket (*b*) has been filled with Dupont plastic lead. This shows that the slope of the cartilaginous roof of the acetabulum corresponds to the bony component above, which is utilized in determining the acetabular index. The floor of the hip socket lies laterad to the lateral accented line of Köhler's tear-drop. The position of the cartilaginous labrum is shown at *a*. It will be seen that this clasps only one-third of the bulbous femoral head, which has been outlined by air. The antero-inferior iliac spine is indicated by the upper arrow.

1. The acetabulum at birth is relatively shallow, due to a thick cartilaginous floor which lies considerably laterad to the lateral accented line of Köhler's "tear-drop."

2. The ligamentum teres is strongly developed, and represents a very real anatomical structure.

3. The femoral head, which is cartilaginous, is large and bulbous, and is apparently enclosed by only a small portion of the cotyloid ligament at birth (one-third only being clasped by the labrum at this time) (Fig. 3).

If these facts are borne in mind, the roentgenologist will avoid an erroneous diagnosis of a hypoplastic capital epiphysis, since what he actually sees is a small bony

the femur to the acetabulum. In addition, slight variation in soft-tissue detail in an otherwise normal-appearing hip may point to the presence of infection. Timely recognition of such diseases as coxitis and osteochondropathy may mean the difference between a satisfactory end-result and subsequent crippling deformity (15, 27, 65). Furthermore, many authorities stress the importance of early diagnosis in congenital dislocation of the hip (22, 28, 41, 60, 61, 74). Since therapeutic efforts after the third year of life are too often only palliative and prompt treatment not infrequently results in a sound, well functioning hip, early and accurate roentgen diagnosis is imperative.

For purposes of description, diseases of



Fig. 4. Case 1. A. Bilateral dorsal dislocation of the hips. Shenton's line is abnormal. Bilabiation of the ilium is seen at *b*. The bony portions of the femoral heads appear hypoplastic. The slope of each acetabular roof is relatively marked.

B. Appearance of the hip joints one year later, following bloodless reduction. The angle of acclivity is still quite marked, the femoral heads are in relatively good position. The left femoral head tends to subluxate slightly laterad. See also Fig. 4, C.

the hip in infants and young children in the Newington Home and Hospital for Crippled Children have been classified in the following manner.

- I. Dislocation
 - A. Congenital
 1. Congenital dislocation
 2. Arthrogryposis
 3. Dislocation associated with or secondary to spina bifida
 - B. Acquired
 1. Traumatic
 2. Septic
 3. Still's disease
 4. Poliomyelitis
 5. Subluxation in muscular dystrophy
- II. Osteochondropathy (Legg-Perthes disease)
- III. Congenital coxa vara and epiphyseolysis
- IV. Rare lesions
 - A. Myositis ossificans
 - B. Achondroplasia
 - C. Osteogenesis imperfecta

I. A: Congenital Dislocation of the Hips

An interesting hypothesis explaining the occurrence of congenital dislocation of the hip was advanced by Le Demany (42) in 1908. In view of his studies in anthropology, comparative anatomy, and em-

bryology, he postulated that in man the large cranium has fostered a wide pelvis to provide an adequate birth canal. The wide pelvis, in turn, engenders long femora for proper balance of the erect individual. Consequently, the human fetus, developing within the ovoid uterus, is gradually forced into an unnatural position. With the lower extremities held in flexion, abduction, and external rotation, pressure exerted upon the knees by the fetal envelopes levers the femoral head from the acetabular fossa, the anterior-inferior iliac spine acting as a fulcrum. This decreases the pressure within the joint, producing a shallow acetabulum. The hip is now, in a manner of speaking, prepared for luxation. This end-point may be reached when the child assumes the erect posture. Due to the effect of muscle pull and body weight, luxation may occur gradually, without evidence of pain. Le Demany is positive in his assertion that "congenital dislocation of the hip" does not occur until after birth, although the stage is set during fetal life.

Two other theories are worthy of consideration: Hart (31) believes that primary

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dislocation is the result of a dysplastic acetabulum, a term indicating a congenital abnormality with hip joint instability and incongruity. According to this authority there is no "dislocation gene" but rather a "hip-dysplasia gene." He emphasizes Putti's (60, 61) statement that dysplastic acetabula are more frequent without dislocation than with it.

On the other hand, Badgley (1) favors the view that hip dysplasia results from abnormal rotation of the limb bud. He believes that marked anteversion of the head results in secondary flattening of the socket, due to the trochanter pressing upon and causing hypoplasia of the posterior acetabular rim. The dysplastic socket fails to retain the femoral head, which migrates posteriorly as the result of gluteal muscle traction. In other words, environment rather than heredity is the determining factor.

In arthrogryposis multiplex congenita, a rare disease characterized by intra-uterine muscle aplasia and joint ankylosis, anterior dislocation of the hip is a relatively common finding. The limbs remain externally rotated. The femoral heads are found to lie ventrally, resting against the iliacus muscle and the anterior margin of the ilium, where a secondary acetabulum is formed. The position of the limb is comparable to that existing in the fetus at approximately the fourth month (1).

Between 1925 and 1944, 77 congenitally dislocated hips were treated at the Newington Home and Hospital for Crippled Children. Of these, 12 were in males and 65 in females. In 22 cases the dislocation was bilateral. The left hip was found to be involved in 19 cases, and the right in 14. Bloodless reduction was affected in 40 cases, and open reduction was required in 37. Crosby (17) in a follow-up study of 28 recent cases found the results of treatment based on clinical and roentgen findings to be as follows: good, 11 cases; fair, 9 cases; poor, 8 cases.

The following case reports have been selected from our files because of their roentgen and clinical interest:



Fig. 4, C. Case 1 (shown also in Fig. 4, A and B). End-result five years after initial treatment, showing excellent reduction of both hips. Minimal sclerosis of the left acetabular roof is noted at *a*. A small sequestrum is shown at the arrow. This is of no roentgen or clinical significance at present. The Trendelenburg test is negative. There is no pain or limp.

CASE 1 (Fig. 4, A, B, C): M. B., white female, age 1 year. Service of Dr. M. M. Pike. *Diagnosis:* Dislocated hips.

Bilateral dislocation was first noted at one year and treated by manipulation on three different occasions. The hips were placed in 90-90° plaster spica for ten months. Recent evaluation of the patient's general condition, six years after first admission, shows a normal range of motion, negative Trendelenburg sign bilaterally, and no pain or limp. In spite of a 3/4-inch shortening of the right leg, the end-result is excellent.

Comment: In this instance diagnosis is readily made on viewing the first roentgenogram (Fig. 4, A). At present the hip sockets appear relatively normal, with only moderate sclerosis of the acetabular roof, which shows a small sequestrum near its outer border (Fig. 4, C). According to Gill (28), all cases of this type should be carefully followed until "perfect restoration of anatomical structure" can be demonstrated by roentgen study. The time interval necessary to effect a perfect end-result may range between one and fifteen years.

CASE 2 (Fig. 5): V. B., age 9 years. *Diagnosis:* Dislocated hips, congenital.



Fig. 5. Case 2. Unsatisfactory result in congenital dislocation following osteotomy of both hips. Four-inch shortening of the left leg. A bilateral positive Trendelenburg sign is elicited. The patient has no pain, but marked limp. Note the acclivity of both acetabula and the marked pelvic tilt. The fragmented, hypoplastic left femoral head is shown at *a*.



Fig. 6. Case 3. Bilateral congenital dislocation, untreated. Bilateral positive Trendelenburg sign and piston motion of both hips. Patient walks and plays without tiring. The femoral heads are hypoplastic and irregular in contour, as shown at *a*. Because the patient is relatively asymptomatic, no therapy is indicated.

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Bilateral dislocation was first recognized at the age of eight. There had been no treatment prior to admission. The patient was the first of five children. The siblings were normal. The child walked at fourteen months. Treatment was as follows: on May 21, 1930, open reduction and osteotomy of right hip; on Jan. 7, 1931, open reduction and osteotomy of left hip. Postoperatively there was evidence of wound infection on the left, with over-riding at the osteotomy site. Both femoral heads were reduced at the time of discharge, Jan. 7, 1932. The final result, seventeen years after admission, is limitation of motion in both hips with 4-inches shortening on the left. A bilateral positive Trendelenburg sign is elicited. There is no pain, but marked limp. Figure 5 shows persistent superior dislocations and marked acclivity of the acetabula, which are malformed and inadequately developed.

Comment: Although it is impossible to state with certainty what the outcome would have been without therapy, the end-result of this case is in sharp contrast to the following untreated case.

CASE 3 (Figs. 6 and 7): W. Z., white female, age 13 years. Service of Dr. C. W. Goff. *Diagnosis:* Dislocated hips, congenital.

This patient was first seen at the age of eleven in September 1945. The greater trochanters were found to lie posteriorly and above the level of the anterior-superior iliac spines. The Trendelenburg sign was positive bilaterally. Piston mobility was present on both sides. There was moderate lordosis, but the leg lengths were equal. At present, two years and two months after her first examination, the patient walks exceptionally well. She can walk and play without tiring and without restriction of activity. The motion of both legs is as follows:

	Right	Normal	Left
Internal rotation	90°	30°	90°
External rotation	50°	60°	50°
Abduction	30°	50°	35°
Adduction	30°	45°	30°
Flexion	90°	60°	100°

No treatment is contemplated for obvious reasons.

The following case presents several characteristic features of arthrogryposis (amyoplasia congenita).

CASE 4 (Fig. 8): C. A., white female, age 15 years. Service of Dr. C. W. Goff. *Diagnosis:* Arthrogryposis.

The patient was admitted at the age of two years with wrists and elbows ankylosed and both hips dislocated. A diagnosis of arthrogryposis was obvious. Treatment consisted of daily physiotherapy, stretching of the adductors, and foot stretching. Both legs were placed in traction at the age of

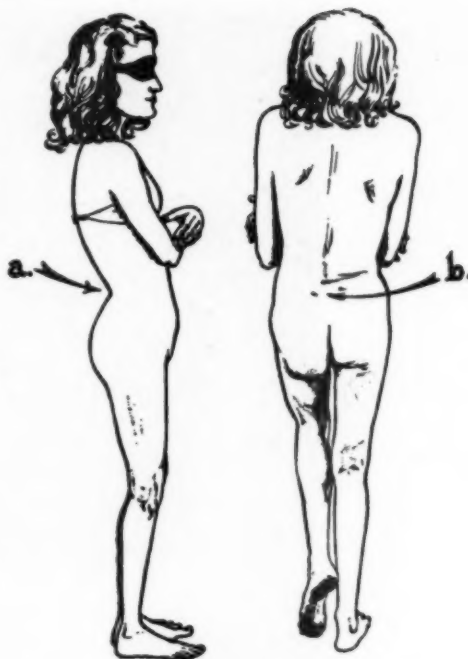


Fig. 7. Case 3. Moderate lumbar lordosis is present at *a*. A positive Trendelenburg sign is observed on the left side at *b*.

seven, with the object of improving the marked flexion deformity of the knees. Multiple operative procedures were performed to correct the elbow, wrist, knee, and foot deformities. The original roentgen appearance is shown in Figure 8, A. During the period of hospitalization, the superior dislocations reduced spontaneously. The exact time and manner of reduction is unknown, although it is well shown in Figure 8, B. Note that the limbs are considerably rotated externally and very satisfactory hip sockets have formed. There is evidence of delayed fusion of the ischial pubic junctions.

Comment: Sheldon (70) states that in arthrogryposis the limbs are usually, though not always, symmetrically involved. Fixation of one or several joints may occur. Of interest is the fact that in fetal arthrogryposis quickening may occur late in pregnancy or be absent altogether. A presumptive intra-uterine diagnosis can be made on the basis of such a history and roentgen demonstration of the fetal extremities fixed in a position of extension. Spontaneous reduction of hip dislocation in arthrogryposis is distinctly unusual,

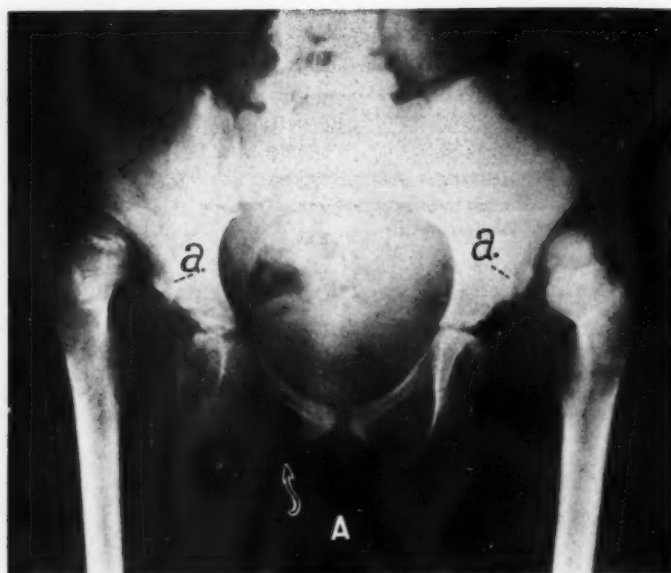


Fig. 8. Case 4. A. Bilateral congenital dislocation in patient with arthrogryposis. Wrists and elbows were ankylosed. Note the high and posterior position of the femora as shown by the bilabiation of the iliac bones at *a*. Spontaneous reduction of the hips occurred as shown in B. Dislocation of the hips in arthrogryposis is usually irreducible.

B. Appearance of the hips following spontaneous reduction. Note the club hand at *a*.

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Fig. 9. Case 5. Dislocation of the right hip in patient with myelodysplasia. Huge spina bifida is shown at *a*. Dislocation of the hip is present at *b*. This may result from muscle weakness with subsequent development of adduction and flexion deformity of the hip (Badgley). Such a dislocation is not necessarily congenital. There is a large defect in the acetabular roof at *b*. This is sometimes referred to by European writers as the *coup d'angle* or nail gouge.

since it has been pointed out that, characteristically, there is anterior and usually irreducible dislocation of the femoral heads (1). Soft-tissue roentgenograms may prove helpful in revealing evidence of aplasia affecting certain muscle groups. Note the obvious deformity of the hand in Fig. 8, B.

In the following case, dislocation of the right hip is associated with a large spinal defect.

CASE 5 (Fig. 9): A. R., age 4 years and 3 months. Service of Dr. R. M. Yergason. *Diagnosis:* Dislocated right hip secondary to huge spina bifida.

A large meningocele was repaired at birth. The child walked normally until the age of four, when he fell and subsequently refused to walk. On ad-

mission a 10° flexion contracture of the right hip was noted. The greater trochanter was found at the level of the iliac crest and posteriorly. Motion was restricted in all directions. Roentgenograms revealed a huge spina bifida, extending from T-10 to S-2, and complete dislocation of the right hip. Treatment consisted of closed reduction on Feb. 11, 1947. A second reduction was required on April 23, 1947, because of redislocation. On June 27, 1947, an abduction splint was applied. On the last examination there was evidence of free motion of the limb at the affected joint. The dislocation has been temporarily corrected.

Comment: Dislocation of the hip with spina bifida has been infrequently described in the roentgen literature. Ingraham (35) finds the incidence of congenital

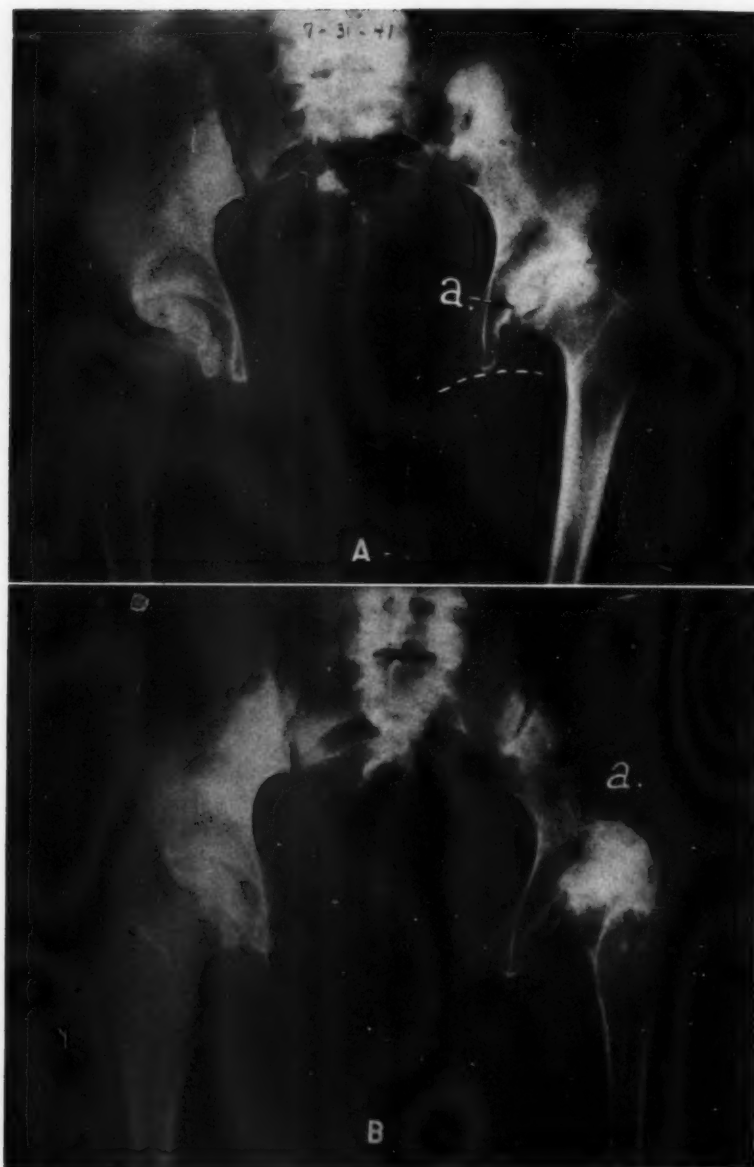


Fig. 10. Case 6. A. Subluxation in patient with non-specific coxitis. Note the sequestra in the region of the fovea and the interruption in Shenton's line. The femoral head tends to luxation in a cephalad direction.

B. Postoperative roentgen appearance following intra- and extra-articular arthrodesis. The adduction deformity, previously present, has been corrected. The cartilage joint space has been completely obliterated (a).

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dislocation associated with imperfect closure of the laminae to be approximately 4 per cent (23 of 570 cases). In the cases reported by Nathanson and Lewitan (52), dislocation was attributed to two factors: (a) relaxation of the ligamentous structures and (b) muscle imbalance. In other words, the pathogenesis of the lesion is somewhat similar to that occurring in poliomyelitis (see Case 8). It is quite possible that in the foregoing case a myelodysplasia accounts for the recurrent dislocation, and that, as the result of the meningocele, normal innervation of the limb bud was prevented (2). However, when simple spina bifida is associated with dislocation, we are inclined to agree with Mayfield (48) that inasmuch as the two conditions occur so often individually, and rarely together, only coincidental significance should be attributed to this finding.

I. B: Acquired Types of Dislocation

The following is a case of infectious coxitis which resulted in luxation of the left hip.

CASE 6 (Fig. 10): G. P., white female, age 15 years. Service of Dr. M. M. Pike. *Diagnosis:* Infectious coxitis with luxation.

This patient had a hemangioma of the cheek excised in 1929 and again in 1932, with implantation of radon seeds. In 1929, lymphadenopathy developed in the "left groin." Following incision and drainage, culture revealed a mixed flora of hemolytic streptococci and *Staphylococcus aureus*. Subsequently, there was a subcutaneous abscess of the left ankle. This was associated with a long period of infection and breakdown of the facial hemangioma.

The patient was first seen at Newington in 1940 because of a flexion and adduction deformity of the left hip. Motion was markedly restricted and there was shortening of the extremity of 1 inch. Intra- and extra-articular fusion was accomplished in October 1941 with the use of tibial and iliac bone. There was no evidence of tuberculosis. The process was felt to be due to an old non-specific coxitis. Roentgenograms show the appearance of the hip before and after arthrodesis.

Comment: Unfortunately, early films of this case are not available. The radiographic appearance is characteristic of an old suppurative coxitis. The clinical course suggests the possibility of a previously un-

recognized pelvic abscess. It should be recalled that, since the capsular lymphatics drain to the external iliac and hypogastric nodes, an iliac abscess may be encountered relatively early with pyoarthrosis of the hip. An abscess not infrequently forms on the ventral surface of the iliopsoas muscle and ultimately infection tends to gravitate to the mesial and upper portion of the thigh (24). For this reason, careful inspection of the pelvic soft parts is necessary whenever a hip lesion is suspected: conversely, every case of pelvic abscess requires close inspection of the adjacent hip joint before a possible suppurative coxitis can be excluded. This type of pelvic lesion should be readily differentiated from the positive obturator sign, which is usually less prominent. The value of the latter sign was first pointed out to us by Drs. E. P. Pendergrass and E. L. Eliason in 1937. More recently, it has been completely described by Hefke and Turner (32), who find that undue prominence of the obturator internus muscle border and obscuration of its margins may be the earliest sign of hip joint inflammatory change. The soft-tissue findings are stressed because they are of distinct aid in early diagnosis, and without their recognition marked impairment of motion and ankylosis may be expected (16, 65).

According to Key and Conwell (38), pathological dislocations are almost always posterior in type and usually result from (a) infantile paralysis, (b) acute synovitis, or (c) chronic destruction of the femoral head or acetabulum. Cumston (13) states that pathologic dislocation is an infrequent sequel to infection but when present is encountered most frequently in the hip joint. The following case histories are illustrative:

CASE 7 (Fig. 11): E. W., white female, age 8 years. Service of Dr. M. M. Pike. *Diagnosis:* Still's disease with dislocation of the left hip.

The diagnosis of Still's disease was made at the age of two. The clinical story was typical of a "juvenile arthritis deformans" with polyarthritis, fever, and enlarged liver and spleen. Roentgenograms showed periarticular soft-tissue swelling of the hands and slight demineralization. The patient was admitted to Newington at the age of six with

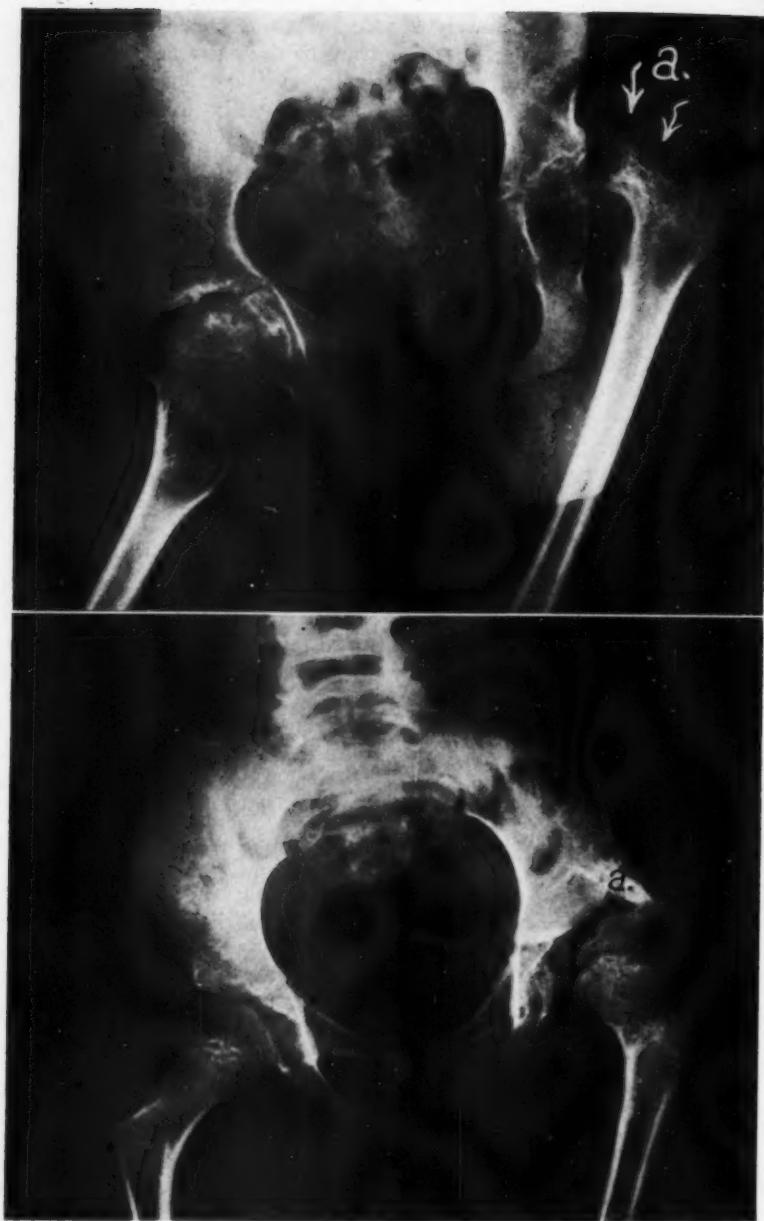


Fig. 11 (above). Case 7. Acquired dislocation in Still's disease. Dislocation of the femoral head is shown at *a*. There is a pronounced adduction and flexion deformity of the femur. Evidently, there has been considerable destruction of the femoral head.

Fig. 12 (below). Case 8. Luxation of the left hip following poliomyelitis. The pelvic girdle and femora are demineralized. The bony shelf at *a* is the result of a previous fascial stripping. This type of dislocation often results from muscle imbalance or habitual posture.

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Fig. 13. Case 9. Bilateral subluxation in patient with spinal muscular atrophy of the Werdnig-Hoffmann type. The muscle bundles are atrophied at *a*, and the pelvic girdle and femora are demineralized. The femora are externally rotated.

the polyarticular swellings of Still's disease. There were obvious hepatomegaly and splenomegaly, fusiform swelling of the fingers, and contractures of the hips, knees, elbows, and wrists. Treatment was conservative because of the activity of the arthritic process. Roentgen study of the hips revealed considerable demineralization of bone and a dislocation of the left hip. The irregular cortical margins of the left femoral head are well shown in Figure 11. Considerable adduction deformity is also evident.

CASE 8 (Fig. 12): N. F., female, age 5 years. Service of Dr. R. M. Yergason. *Diagnosis:* Luxation of left hip following poliomyelitis.

The past history revealed poliomyelitis at six months of age, involving the legs, trunk, and right arm. The patient was admitted to Newington at the age of four with flexion contracture of the left hip and a tight heel cord. Fascial stripping and a Hoke tenotomy were performed. For a year the patient has been ambulatory with two long leg braces and a corset. Recent films show beginning luxation of the left hip and a pelvic tilt to the right.

Comment: Elzinga and Key (20) state that with paralysis of the pelvic musculature hip dislocations may be expected in approximately 2.5 per cent of the cases. In their experience, such dislocations are

more often right- than left-sided, and seldom occur bilaterally. The deformity may result from muscular imbalance or from habitual posture. On the basis of muscle power studies, these authors conclude that "severely paralyzed hips with relatively strong flexors, adductors, and internal rotators and relatively weak extensors, abductors and tensor-fasciae femoris may be expected to dislocate. It is further evident that relatively strong adductors combined with relatively weak abductors and tensor-fasciae femoris are especially likely to result in dislocation." Because of instability resulting from muscle paralysis, hip fusion (30) in such cases is often necessary.

Another cause of pathological dislocation in our experience has been muscular dystrophy. The following is a case in point:

CASE 9 (Fig. 13): W. C., white male, age 12 years. Service of Dr. C. W. Goff. *Diagnosis:* Spinal muscular atrophy, Werdnig-Hoffmann type.

Poor muscular development was first noted at

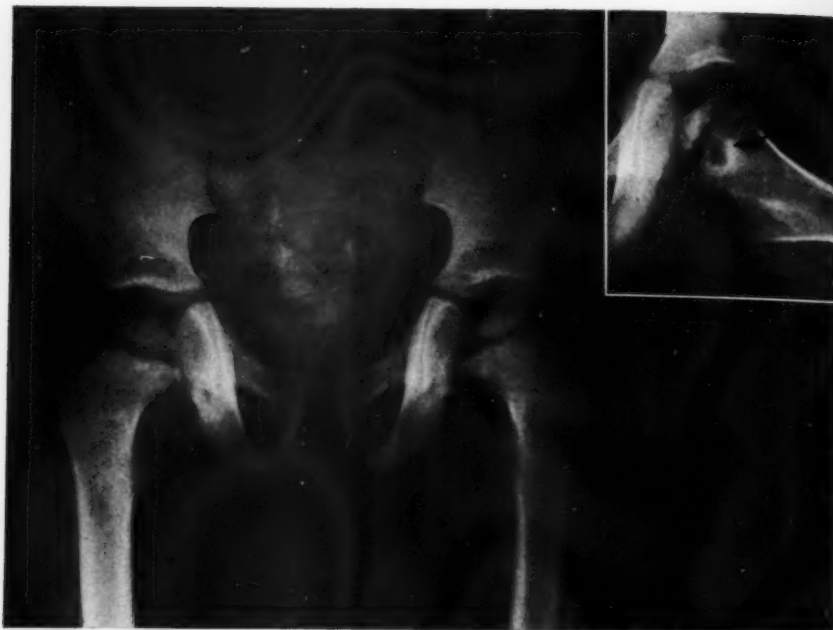


Fig. 14, A. Case 10. Admission roentgenogram showing bilateral Legg-Perthes disease at age of three. There is a definite metaphysitis, which is well shown in the lateral roentgenogram (see insert). The hip joint spaces are definitely widened. See also Fig. 14, B-E.

the age of ten months. The child has never stood or walked without assistance. One brother and one aunt were similarly affected. The patient now leads a wheel-chair existence in a celastex body jacket. Roentgenograms show a subluxation of both hips laterally, marked demineralization of bone, and apparent valgus deformity of the femora (Fig. 13). The muscle bundles of the thigh are markedly atrophied.

Comment: The Werdnig-Hoffmann infantile type of spinal muscular atrophy is said to be due to degeneration or loss of the motor cells of the spinal gray matter, particularly in the lumbosacral region (23). Clinically, the weakness usually begins in the gluteal and pelvic muscles and then spreads to involve the thighs and shoulder girdles. In the roentgenogram, as on physical examination, the muscles and soft tissues can hardly be distinguished from each other.

II. Legg-Perthes Disease (Osteochondritis Deformans Juvenilis)

Legg's original description of "an obscure affection of the hip joint," based on clinical

and roentgen observations in five cases (43), is surprisingly accurate. In his second case, for instance, he reported "irregular areas of increased radiability near the epiphyseal line." Thus attention was first directed to the metaphysitis which Gill (27) has shown us more recently is such a relatively constant feature of the disease.² Legg stated: "It is worthy of note that all these cases sought advice solely on account of the limp." In 1918, he reported further experience with 75 personally observed cases (44), concluding that "flattening of the upper femoral epiphysis is due to a circulatory disturbance at the epiphyseal line, causing atrophy and flattening . . ."

Gall and Bennett (25), in a very complete treatise, report autopsy findings in the case of a thirteen year-old boy with positive roentgen findings of Legg's disease. They found the femoral neck widened and its long diameter shortened.

² Doub (19) has pointed out that the metaphysitis and the lesion of the capital epiphysis (coxa plana) usually occur simultaneously.

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Fig. 14, B-E. Case 10 (shown also in Fig. 14, A). Bilateral Legg-Perthes disease. B. Film made eight months after that reproduced in Fig. 14, A, showing definite progression of the disease on the left side and aseptic necrosis of the femoral heads.

C. The capital epiphyses show further evidence of degeneration one year later. Marked flattening of the left capital epiphysis is present at *a*. There is a bilateral positive Waldenström's sign.

D. Improved status of both capital epiphyses during the stage of recovery. This stage lasts 24 to 36 months, more or less. The left capital epiphysis is relatively granular, though definite regeneration is evident.

E. Final result after prolonged immobilization in a Wu-splint. Note the relatively normal hip joints in spite of the severe osteochondropathy shown on the original roentgenogram (Fig. 14, A).

In the femoral head there was evidence of fibrillation of the matrix of the cartilage cells, imperfect alignment of these cells, and invasion of the marrow spaces by fibrous connective tissue. The vessels of the ligamentum teres were found to be intact.

Pike (55), in a practical article, states that the lesion may involve either the head or neck of the femur, or the acetabulum.

The greatest incidence is during the stage of active development, that is between the ages of five and ten years. The classical findings suggest a mild inflammatory disturbance. Since operation or biopsy is rarely indicated, Pike places considerable reliance on early roentgen diagnosis as exemplified by (*a*) mottled areas of increased and decreased density in the capital epiphysis, (*b*) fragmentation, and (*c*) widen-

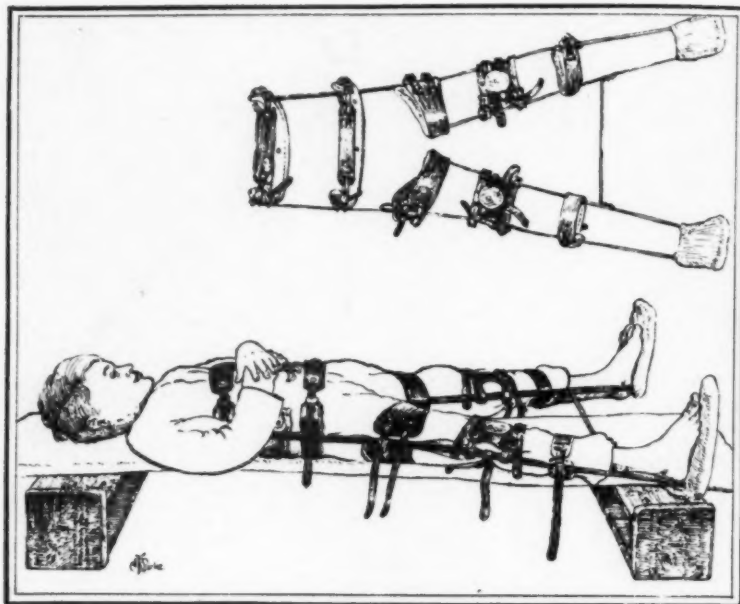


Fig. 15. Illustration showing Wu-splint as used at the Newington Home and Hospital for Crippled Children. This is a simple orthopedic device which prevents the action of muscle spasm and gravity from affecting the plastic proximal femoral epiphysis in Legg-Perthes disease.

ing of the cartilage space (Waldenström's sign).

The disease can be divided roughly into two stages, which may merge imperceptibly into each other. They are the *stage of onset and activity*, which is stated to last eighteen months, more or less, depending upon the accuracy of the history and the type of treatment employed, and the *stage of recovery*, which is said to last twenty-four to thirty-six months, more or less.

Pike has also reported autopsy findings of Legg's disease in a five-year-old boy who was severely injured in an automobile accident and died seventy-two days after admission to the Hartford Hospital. Microscopic studies showed evidence of bone infarction, hemorrhage, necrosis, and mild inflammatory changes. Through the courtesy of Dr. John H. T. Sweet, the sections from this case have been recently reviewed (Fig. 16). We are indebted to Dr. Perry Hough, assistant pathologist at the Hartford Hospital, for the following description.

Gross: Longitudinal section through the head of the left femur shows some flattening of the surface and an irregular area of degeneration between the articular surface and the epiphyseal line. Below this there is an area of pale yellow color replacing the normal red marrow.

Microscopic: Section through the head of the femur shows an essentially normal articular cartilage. The bone between this and the epiphyseal cartilage shows an extensive and rather sharply demarcated area of complete necrosis consisting of amorphous debris and necrotic bony trabeculae. Through this area there is no cellular infiltration or inflammatory reaction. Adjacent to this region there is active bone production. The marrow spaces contain a loose, rather vascular granulation tissue through which there is proliferating fibrous connective tissue. The epiphyseal cartilage appears normal. Distal to this there is a sharply demarcated area in which all of the normal hematopoietic elements have disappeared, leaving only a fatty marrow. These changes have the appearance of aseptic necrosis due to infarction of bone.

In the twelve-year period between 1935 and 1947, 59 cases of osteochondropathy of the hip (Legg-Perthes disease) have been treated at the Newington Home and Hospital for Crippled Children. The average

age on admission was eight years. In this group there were 47 male patients and 12 females. Ten bilateral cases were noted. Physical examination revealed limp and limitation of motion in all cases. Of the 59 patients, 54 complained of pain, which was not infrequently referred to the knee. Shortening was demonstrated in 13, and atrophy in 16.

The radiographic findings were quite characteristic. Fragmentation of the femoral head was present in all cases, associated with a minor or marked degree of coxa plana. Broadening of the femoral head was observed in 44 cases, and migration of the head in 13 instances. Such migration, according to Pike (55), results from improper protection of the femoral head at a time when it is still relatively plastic. One explanation of the manner in which the forces of strain and stress produce such a migration has been advanced by Jansen (36), who believes, with others, that Legg-Perthes disease is due to a dysplastic acetabulum.

CASE 10 (Fig. 14, A-E): N. J., white male, age 8 years. Service of Dr. M. M. Pike. *Diagnosis:* Legg-Perthes disease, bilateral.

The patient was a normal healthy child who began to limp and complain of pain in the left knee at the age of three years, three months prior to admission. There was no injury antedating the symptoms.

The child appeared well developed on examination, with a limp and flexor and adductor spasm in the left hip. He was placed in Wu-splint (Fig. 15) recumbency from June 1942 to June 1947, a period of five years. Spasm and restriction of motion disappeared shortly after immobilization. Thereafter the patient was removed from the splint twice weekly for physiotherapy and non-weight-bearing exercises in the pool.

The treatment resulted in practically normal structural regeneration of the epiphyses, full range of motion bilaterally, and equal leg length.

The roentgenograms reveal the various phases of the disease over a five-year period. The stages of bone absorption, regeneration, and complete healing are demonstrated. This case serves to emphasize the importance of the Wu-splint in obtaining a good end-result.

CASE 11 (Fig. 17): F. F., white male, age 9 years. Service of Dr. M. M. Pike. *Diagnosis:* Legg-Perthes disease.

This patient first complained of pain in the left

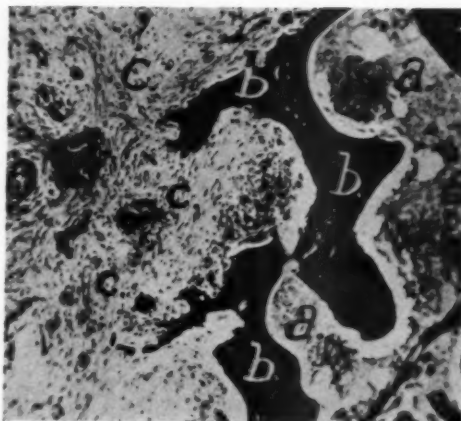


Fig. 16. Microscopic section showing bone infarction in Legg-Perthes disease. Note amorphous debris at *a*. Necrotic trabeculae are present at *b*. The marrow spaces contain vascular granulations at *c*. There is no evidence of cellular infiltrate or inflammatory reaction. Case previously reported in detail by Pike.

hip, associated with limp, in 1944, three years prior to admission. He was treated at home with bed rest for one year, non-weight-bearing splint for one year, and crutch ambulation without weight bearing for one year.

On admission he showed 3/4-inch shortening of the left leg, associated with 1 1/4-inch atrophy of the left thigh and calf. Internal rotation, flexion, and abduction were limited to 15°.

Roentgenograms on admission to Newington, in March 1947, revealed extensive flattening and fragmentation of the left capital epiphysis with widening of the neck and acetabular dysplasia. This picture is frequently seen in individuals not receiving early and completely enforced immobilization.

CASE 12 (Fig. 18, A and B): W. T. Service of Dr. R. M. Yergason. *Diagnosis:* Unusual osteochondropathy associated with intra-articular calcium deposition.

This patient was first seen at the Newington Home at the age of nine. He had been complaining of pain in the right hip for one year. The history revealed the child was born after a normal pregnancy. Nothing unusual was noted until the age of seven weeks, when swelling of the left wrist was observed, followed by swelling of both knees. Roentgenograms at this time revealed marked fragmentation of the epiphyses of both knees and the left wrist, with calcific deposits throughout the joints. Subsequent films at the age of four showed flattening of the femoral condyles due to an epiphysitis (Fig. 18, B).

The patient had received vitamin D in the form of viosterol from five weeks of age. Repeated blood calcium, phosphorus, and non-protein nitrogen determinations were normal.



Fig. 17. Case 11. Indifferent result in Legg-Perthes disease after treatment at home with bed rest for one year, non-weight-bearing splint for one year, and crutch ambulation without weight-bearing for one year. Note the extensive flattening and fragmentation of the left capital epiphysis with widening of the neck and acetabular dysplasia. The changes are well shown at *a*.

Examination on admission to Newington showed 7/8-inch shortening of the right leg, with moderate muscle atrophy and restricted motion of the right hip (Fig. 18A). Treatment was by recumbency in a Wu-splint for two years with subsequent gradual improvement in the radiologic appearance of the joints. The right femoral epiphysis showed good regeneration. The clinical result was 1-inch shortening of the leg, with only slight restriction of abduction and internal rotation.

CASE 13 (Fig. 19, A, B, C): G. R., age 3 years, 9 months. Service of Dr. R. M. Yergason. *Diagnosis:* Osteochondritis of the femoral metaphysis.

This patient was admitted to Newington in September 1944 with a left leg limp of one month duration. Past history was negative except for pertussis at one month. Examination showed slight limitation of abduction and rotation in the left hip. Laboratory findings were as follows: tuberculin test negative; urinalysis negative; Mazzini test negative; sedimentation rate, 9 mm in eleven hours; white blood cells 12,000 (46 per cent polymorphonuclears); blood calcium 10.6 mg.; phosphate 4.4; cholesterolin 191 mg.

Treatment consisted of bed rest. In February 1945, no limitation of motion was noted. The leg lengths were equal. In January 1947, a Wu-splint

was applied to restrain activity. A full range of motion was noted. In April 1947, the patient was allowed up on crutches. In July 1947, he was returned to Wu-splint recumbency because of further destruction at the epiphyseal line.

Roentgenograms show a progressive widening of the epiphyseal line and cystic changes, with no alteration in the bony texture of the epiphysis itself.

Comment: This is an unusual case of osteochondritis of the femoral neck without involvement of the proximal epiphysis, or subsequent appearance of coxa vara deformity.

III. Congenital Coxa Vara and Epiphyseolysis

Congenital coxa vara, first described by Hofmeister in 1894, often presents a picture of hypoplasia of the femoral head and neck. Numerous abnormalities have been described, ranging from complete absence to complete ossification of the head and neck of the femur, with or without fissure formation. Noble and Hauser (53) state that



Fig. 18, A. Case 12. Unusual osteochondropathy associated with intra-articular calcium deposits beginning at the age of seven weeks. Note the marked coxa plana involving the right hip, with roentgen evidence of impaired blood supply to the femoral epiphysis. The patient was treated in Wu-splint recumbency for two years, with subsequent gradual improvement in the radiologic appearance of the joints. See also Fig. 18, B.



Fig. 18, B. Case 12 (see also Fig. 18, A). At the age of four this patient showed evidence of bilateral lesions involving the femoral condyles with flattening of the condyles and obliteration of the intercondyloid notches. These changes have persisted to date although the intra-articular calcium deposits are no

the disease appears during the first four years of life, after which ossification of the neck is complete. Various etiologic factors have been suggested. The most acceptable is Reiner's hypothesis that a vascular disturbance upsets the calcification of the femoral neck and epiphyseal line.

The patient walks with a limp or waddle. Deformity takes place gradually without the period of immobility seen in epiphyseolysis. Usually limitation of abduction and flexion deformity are present. The gait is cross-legged in bilateral cases. Prominence of the trochanter and lordosis of the back are marked. There is shortening of the extremity and the Trendelenburg sign is positive.

longer present. This may be termed a "chondrodystrophia calcificans congenita," although we prefer to consider it as a widespread manifestation of osteochondropathy associated with intra-articular calcium deposits.



Fig. 19. Case 13. A. Osteochondritis of the left femoral neck on admission. Note the widening of the femoral neck and definite roentgen evidence of metaphysitis as shown in the insert.

B. A year later there has been further slight progression of the lesion without roentgen evidence of coxa vara or coxa plana deformity. See also Fig. 19, C.

The roentgen findings depend upon the extent of the disease. The femoral neck often forms an angle or 90° or less with the shaft. The head may be absent, hypo-

plastic, or normal. An inverted Y-shaped defect consisting of osteoid tissue is sometimes seen in the femoral neck. There may be hypoplasia of the hemipelvis on the

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Fig. 19, C. Case 13 (shown also in Fig. 19, A and B). Further progression of the osteochondritis of the femoral neck three years after original admission.

affected side. The acclivity of the acetabular fossa may be increased. The femoral shaft is often slender.

CASE 14 (Fig. 20): J. C., colored male, age 7 years. Service of Dr. M. M. Pike. *Diagnosis:* Congenital coxa vara.

The patient was admitted complaining of pain and limp of eight months duration, with no previous history of trauma or infection. He walked with a left leg limp. His gait resembled that in congenital dislocation of the hips. There was a 3/4-inch shortening of the left leg with a tilt of the pelvis to the corresponding side. Abduction and internal and external rotation were limited. The Trendelenburg sign was negative, and piston-mobility of the hip joint was lacking. The laboratory findings were negative.

Treatment consisted of preliminary traction to overcome muscle spasm, followed by a subtrochanteric osteotomy of the left side. No internal fixation was employed. Roentgenograms (Fig. 20) showed a typical congenital coxa vara. The varus deformity amounts to 90° on the left and 123° on the right side. The granular appearance of the left metaphysis and the tendency to subluxation of the epiphysis are worthy of note.

Slipped femoral epiphysis, or adolescent coxa vara, is encountered most often be-

tween the ages of twelve and twenty-one. It is more frequently seen in males, three to one. Early diagnosis is important, inasmuch as the end-results are somewhat better than in Legg-Perthes disease.

The onset of the condition is insidious. Painless limp and external rotation are followed in time by dull pain. In early stages of the disease roentgenograms appear negative unless subjected to the closest scrutiny.

The most widely accepted etiologic factor is an endocrine disturbance, one-third of the cases presenting a picture of dystrophia adiposogenitalis. Hypofunction as well as hyperfunction of the anterior lobe of the pituitary is said to produce metaphyseal weakening. Brailsford (6, 7) states that renal rickets is an important cause. Milch (51) feels that the counterforce exerted by the iliopsoas tendon against the externally rotating femoral head plays an important etiologic role.

The term "preslipping stage" is a misnomer (50, 51). Early diagnosis requires

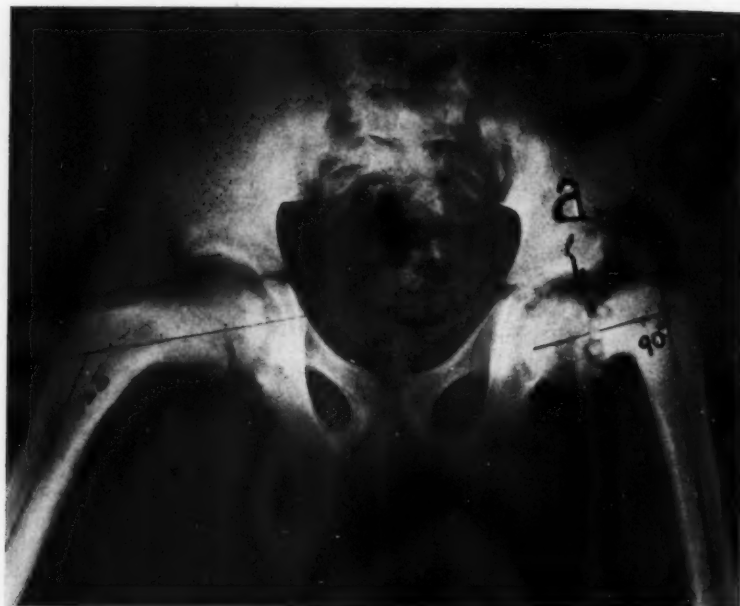


Fig. 20. Case 14. Typical coxa vara deformity involving both hips. The varus deformity on the left amounts to 90° ; on the right to 123° .

close attention to slight variations in contour of the femoral head and adjacent neck. After the tenth year of life the femoral head develops more rapidly than the neck and projects above it, producing the so-called "capital hump" (Pomeranz, 57). Rotation of the femoral head through multiple obliquities will not obliterate this hump nor alter the height of the head. The lower border of the femoral neck is normally concave. Convexity should immediately suggest a possible coxa vara.

Loss of the capital hump, best demonstrated in lateral projections, is the earliest diagnostic sign of epiphyseolysis. As the neck rotates forward on the capital epiphysis, a beak-like prominence of the lower border of the head appears. Finally, as the head and the neck separate, cleavage occurs anteriorly. This forms a V-shaped hiatus, creating the illusion of a widened epiphyseal line. Another helpful diagnostic point is the reduction of the height of the femoral head, as observed in the anteroposterior roentgenograms. This indicates posterior rotation of the head. In

the later stages of slipping, three crescentic shadows appear. These represent the anterior rim of the femoral neck and the posterior and anterior margins of the femoral head (Pomeranz and Milch, 50, 57). Old or unrecognized cases present a picture of a mushroom-like femoral head with eburnation of the articulating surfaces, coxa vara, and frequently osteoarthritic changes involving the acetabulum.

CASE 15 (Fig. 21, A, B, C): I. T., white female, age 12 years. Service of Dr. R. M. Yergason. *Diagnosis:* Epiphyseolysis.

The patient was admitted April 8, 1947, with pain in the right hip of five months duration. There was no history of previous trauma. The family history was not relevant. One month prior to admission closed manipulation and immobilization in a spica cast (elsewhere) were unsuccessful.

The patient was well developed, with a pronounced right-sided limp. There was limitation of flexion, internal rotation, and abduction. The right leg was $1/2$ inch shorter than the left and showed $3/4$ -inch atrophy of the thigh and calf. Considerable muscle spasm was noted.

Treatment consisted of open reduction with wedge-osteotomy of the femoral neck and fixation with a Lloyd screw. Full range of motion was re-

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Fig. 21. Case 15. A. Epiphyseolysis showing loss of capital hump at *a*. B. Lateral projection of epiphyseolysis, showing V-shaped hiatus at *a* in the semilateral projection. This V-shaped defect may occasionally simulate an epiphysitis when viewed in the anteroposterior roentgenogram. See also Fig. 21, C.

gained one month postoperatively, and the patient started non-weight-bearing on crutches.

Comment: Preoperative roentgenograms show the value of the lateral projection.

The V-shaped hiatus is well shown at "a" in the semilateral projection (Fig. 21, B). The loss of the capital hump is obvious in the anteroposterior view. One of the



Fig. 21, C. Case 15 (shown also in Fig. 21, A and B). Satisfactory end-result following osteotomy and insertion of a Lloyd screw. There is minimal posterior displacement of the femoral head as shown by the decreased height of the capital epiphysis at *a*. Previous closed manipulation in this case had resulted in a complete failure.

crescentic lines described by Pomeranz is clearly demonstrated.

The roentgenograms made postoperatively show the deformity largely corrected. The height of the femoral head, however, remains subnormal and indicates that there is still some anteversion of the femoral neck. We have seen 14 similar cases during the last eight years.

IV. Rare Lesions: A: *Myositis ossificans*

CASE 16 (Fig. 22, A, B, C): M. McG., white female, age 8 years. Service of Dr. C. W. Goff. *Diagnosis:* Bilateral myositis ossificans.

This 8-year-old girl received extensive second-degree burns of the back, arms, and lower extremities in August 1940 and was admitted to another hospital. Her course was stormy, but she was finally discharged July 1, 1941. During her initial hospitalization she received six transfusions, three skin grafts, and required manipulation under anesthesia for contractures of both hips. She was admitted to the Newington Home and Hospital for Crippled Children in August 1941 with flexion con-

tractures of both hips. Range of motion was as follows:

	Right Hip	Left Hip
Flexion	50°	40°
Abduction	5-10°	5-10°
Rotation	5°	5°

Blood calcium, phosphorus and phosphatase studies were all within normal limits.

The diagnosis was bilateral ossification of the iliacus muscle.

Treatment consisted of traction on a Schwartz frame and physiotherapy. In March 1942, a bony bridge extending from the anterior rim of the acetabulum to the femoral shaft was excised from the left hip. A segment of this bridge formed part of the capsule. In April 1942, a similar bony bridge was excised from the right hip, and in December 1946, an additional excision was necessary on the right side.

At present the patient has 60° of flexion bilaterally, 45° of abduction, and full extension. She walks without support and without a noticeable limp.

Radiographic studies in the anteroposterior and semilateral projections show myositis ossificans of the iliacus muscles before and after excision. The dense bony struts are particularly well shown in the roentgenograms dated Nov. 13, 1941 (Fig. 22, A and B.) In the postoperative study, osteophytic spurs in the region of the anterior-superior and inferior iliac spines are noteworthy.

Comment: Extensive myositis ossificans was a frequent finding in paraplegic soldiers in World War II. Soule (72), reporting 62 cases of disease or injury of the spinal cord and cauda equina, observed ectopic deposits in the soft tissues chiefly around the hips and knees. Similar findings were noted in a large group of paraplegics at Percy Jones General Hospital. It was noted that the patients who came under observation had been in dorsal recumbency for many months and frequently presented large decubitus ulcers in the trochanteric areas. We are indebted to Dr. Francis M. McKeever for directing our attention to the importance of chronic infection, spinal cord lesions, and prolonged dorsal recumbency as factors in producing such heterotopic ossification. In the case recorded above, the lesions resulted inadvertently from too vigorous manipulation of the hip contractures while the patient was under anesthesia.

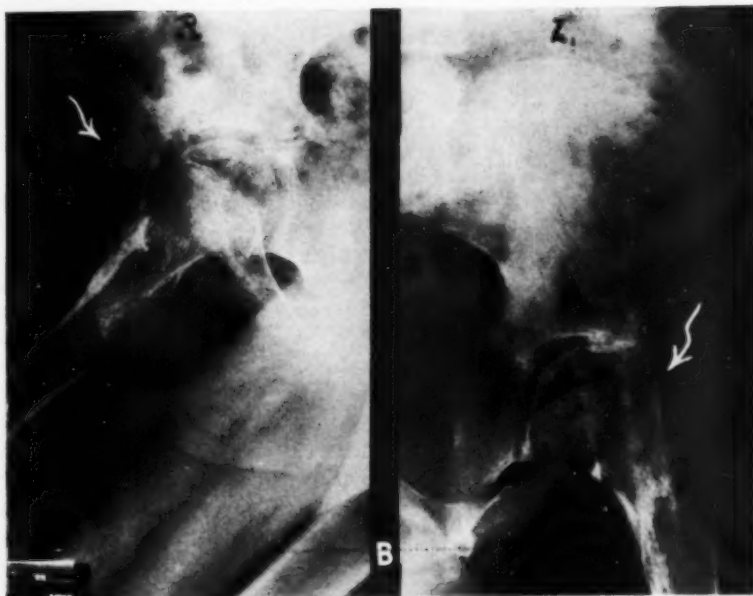


Fig. 22. Case 16. A. Bilateral myositis ossificans following forcible manipulation under anesthesia at another hospital. The broad bony bridges are particularly well shown at *a* and at arrows.

B. Semilateral projection. The ossification corresponds to the course of the iliacus muscle and its tendon. At operation the ossification was found to involve the hip joint capsules bilaterally. See also Fig. 22, C.



Fig. 22, C. Case 16 (shown also in Fig. 22, A and B). End-result after successful removal of bony bridges. There are a number of osteophytic spurs in the region of the anterior-superior and inferior iliac spines.

IV. B. Achondroplasia

CASE 17 (Fig. 23, A, B, C): H. K., female, age 13 years. Service of Dr. M. M. Pike. *Diagnosis:* Achondroplasia, hyperplastic type.

A 13-year-old female dwarf was first seen in the out-patient clinic in 1943 and was admitted to the hospital in 1947. At the age of two the mother noted that the child did not walk and attributed this to the fact that her feet were small. At the age of four, a pediatrician made a clinical diagnosis of achondroplasia. The child's general health had always been good. She was mentally alert and intelligent. There was no antecedent history of dwarfism in the family.

Physical examination showed the height to be 36 inches, with a sitting height of 26 inches. The head was normal except for a receding lower jaw and aclasia of the teeth. The trunk was somewhat shorter than normal, and the extremities were disproportionately short. The anteroposterior diameter of the chest was decreased. The buttocks were prominent. The right leg measured 13 1/4 inches, the left 13 3/4 inches. Each upper extremity, from the acromion to the radial styloid, measured 10 1/4 inches. The patient walked with a waddling gait and tired easily. There was no complaint of pain. Routine laboratory findings were normal.

Roentgenograms (Fig. 23, B and C) show fragmented, hypoplastic femoral epiphyses and broadened femoral shafts. The pelvis is contracted and somewhat triradiate in appearance. The reduced anteroposterior pelvic diameter accounts for the frequency of cesarean section in such cases.

Comment: It has been stated that achondroplastic dwarfs existed 5000 years B.C. (10). They were employed as jesters, acrobats, and animal keepers. The designing and guarding of precious stones and jewels was another occupation reserved for these individuals.

Kaufmann describes three different pathologic types: (a) the *hypoplastic form*, in which the metaphyses show a uniform reduction of cartilaginous proliferation; (b) the *hyperplastic type*, with mushroom-like terminal bony segments overhanging the mid segments of the shafts; (c) a rare or malacic type characterized by softening of the cartilage. Caffey (11) apparently believes that Morquio's disease is probably a variant of hyperplastic or possibly malacic chondrodystrophy, in which disabling deformities are conspicuous.

Bromer (8), on the other hand, feels that the clinical picture as described by Morquio, Ruggles, Meyer, and Brennehan is so different from the usual appearance in achondroplasia that the term Morquio's disease or osteochondrodystrophy is warranted. He points out, however, that most of the cases which we see do not run true to

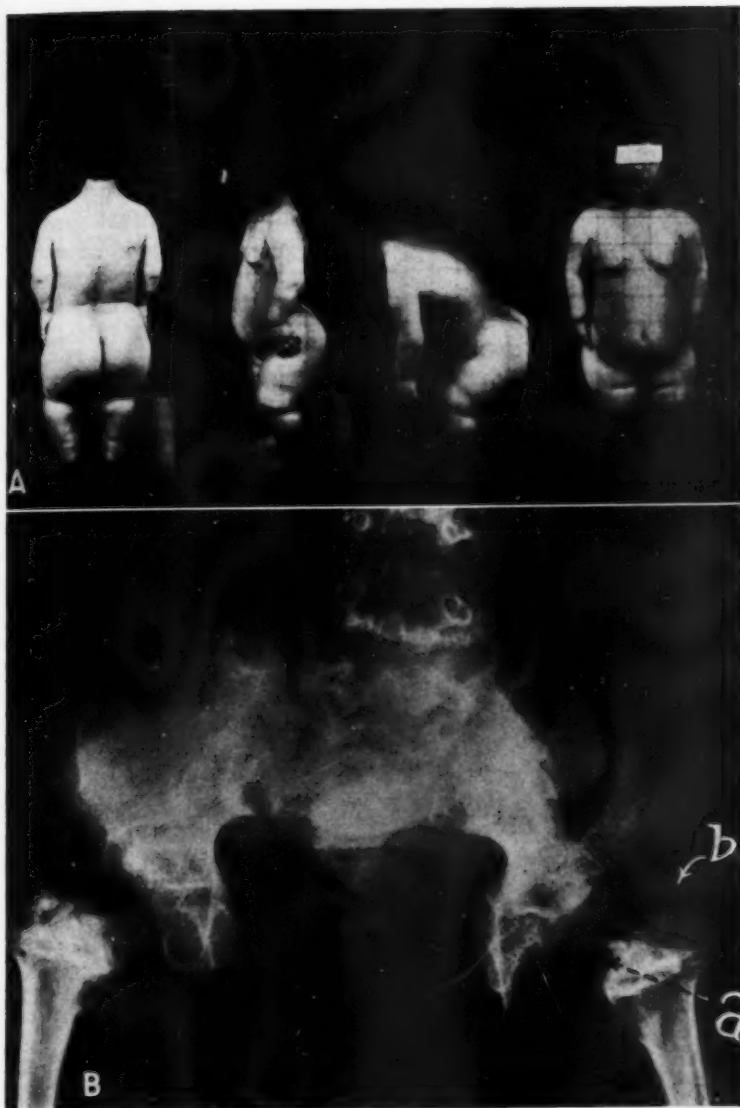


Fig. 23. Case 17. A. Clinical appearance of achondroplastic dwarf, showing the disproportionately short extremities as compared with the trunk. Note the characteristic pot-belly and prominent buttocks.

B. Pelvic girdle in achondroplastic dwarf. This has been classified as the hyperplastic type of achondroplasia. Note the small femoral heads at *a* and the hyperplastic trochanters at *b*. The anteroposterior diameter of the pelvis is reduced. See also Fig. 23, C.

Morquio's description, and are variants or transition forms—*formes frustes*. This disease should not be confused with multiple cartilaginous exostoses, hereditary deforming chondrodysplasia, and diaphyseal ac-

lasis, which are all one and the same: exostoses with an enchondromatous base.

IV. C: *Osteogenesis Imperfecta*

This disease was first described by Lob-

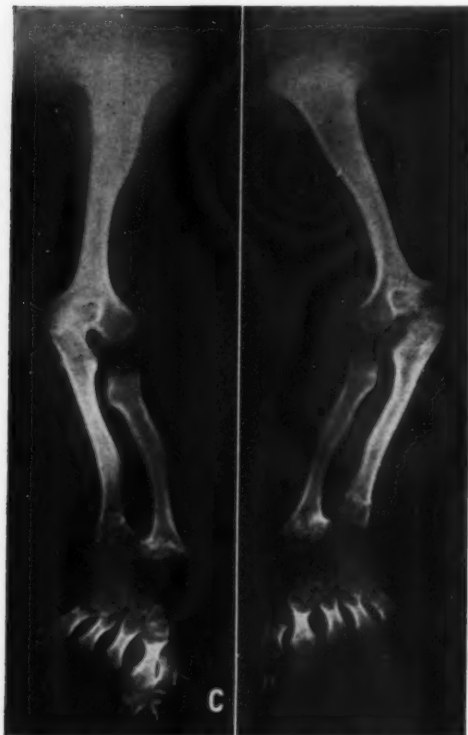


Fig. 23, C. Case 17 (shown also in Fig. 23, A and B). Characteristic appearance of the long bones in patient with achondroplasia. Note the trident configuration of the hands.

stein in 1833, and is characterized by the triad of blue sclerae, brittle bones, and otosclerosis. The three features are not present in all cases, and the blue sclerae may exist with normal skeletal structures, which show no tendency to fracture.

Classification of the entity in the literature is confusing. Many names are used to describe the same condition. Ghormley *et al.* (5) divide their cases into (1) a hereditary (Mendelian dominant) type and (2) a congenital type which may either be present at birth (osteogenesis imperfecta congenita) or appear later in life (osteogenesis imperfecta tarda).

The etiology is obscure. There is probably an inherent germ-plasm defect, with failure of osteoblasts to lay down bone matrix. The parathyroids are never involved. The pituitary, thymus, and thy-

roid all show normal structure on microscopic examination. Blood calcium and phosphorus are normal, and elevation of phosphatase occurs only after fracture.

The bones are demineralized, with thin cortices and expanded metaphyses. Trabeculation is poor: osteoid tissue is increased. The patients tend to be physically frail and small of stature, with large heads and prominent frontal and occipital bones. There is flaring of the lower ribs. The vertebrae are typically biconcave in appearance. A prominent feature is the congenital laxity of the ligaments with hyperextensibility of all joints. The otosclerosis is due to an ankylosis of the stapedo-vestibular joint. The blue sclerae result from thinness and transparency of the connective-tissue layers of these structures.

Various forms of therapy have been tried: pituitary, thyroid, thymic, and ovarian extracts. Ropes (64) has reported encouraging clinical improvement and increased calcium retention in eight cases following prolonged estrogen therapy. Gardner (26), at the Yale School of Medicine, has demonstrated striking endosteal bone formation in immature mice treated with estrogens. We have observed increased calcium retention in one case and microscopic improvement in bone structure in another following estrogen therapy.

CASE 18 (Fig. 24): R. B. White male, age 4 years. Service of Dr. R. M. Yergason. *Diagnosis:* Osteogenesis imperfecta.

This child was born with blue sclerae and a fracture of the left tibia. There was no family history of brittle bones or blue sclerae. Diet and vitamin intake were always adequate. At fifteen days of age the left tibia was refractured, and in the subsequent three and a half years the child sustained 22 fractures, all involving the tibiae and femora. In addition to the deformity and demineralization of the lower extremities, there were flaring of the lower ribs, a tam-o'-shanter skull, biconcave vertebrae, and hypermotility of all the joints, associated with laxity of the ligamentous structures. Blood chemistry, including determinations of calcium, phosphorus, non-protein nitrogen (17), ketosteroids, and estrogens were all normal. Alkaline phosphatase was temporarily elevated following fractures.

Treatment consisted of suspended traction, a trial of antuitrin-S without demonstrable effect, and es-

trogenic therapy consisting of 500 rat units of estrin twice weekly. On the latter therapy calcium retention has increased but there has been no change in bone line density in our roentgenograms.

Roentgen study shows marked demineralization, concentric atrophy of the femoral shafts, and bilateral coxa vara deformity (Fig. 24). Note the exuberant callus formation about the fracture sites.

ROENTGEN TECHNIC

The need for exact roentgen technic is emphasized by recent articles describing such entities as "ischium varum" or "varus or valgus pelvis." It seems clear that in some of these cases an erroneous diagnosis was rendered as a result of faulty positioning of the patient, usually with the pelvis rotated into a slight obliquity. Obtaining a film properly centered and symmetrically exposed in the erect or horizontal posture would have obviated this difficulty. The constant reference to valgus deformity of the femur suggests that in many cases the leg is externally rotated, with consequent foreshortening of the proximal femur, and that in all probability no true valgus was present. It is highly important, therefore, that for anteroposterior roentgenograms the legs be internally rotated, the heels separated one or two inches. Upright films are indicated, wherever possible, in children suspected of having dysplastic acetabula or congenital dislocation. It is surprising how often satisfactory erect roentgenograms can be obtained if an immobilizing band is used. In infants, where this is impossible, an attempt is made to show subluxation by adducting the thighs and exerting pressure in a cephalad direction. Where obvious deformity of the thigh-folds exists, a soft-tissue study is desirable for permanent record even though no dislocation is evident.

We are in agreement with de Lorimier (18) that frequently undue emphasis is placed upon the use of low-kilovoltage technics, and too much contrast results. With somewhat heavier penetration, a more diagnostic film is secured, and lesions become discernible which would otherwise pass unnoticed. This is particularly true of Legg-Perthes disease. For this reason Gill

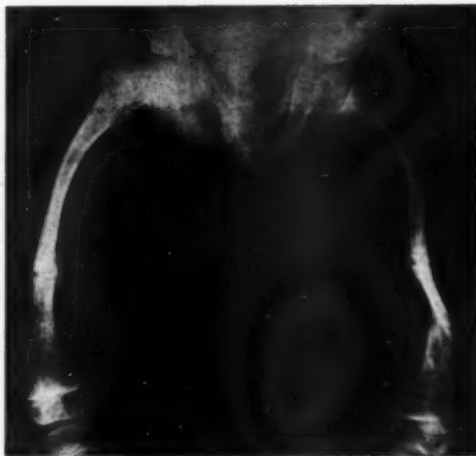


Fig. 24. Case 18. Coxa vara deformity in patient with osteogenesis imperfecta. Multiple healing fractures are seen.

(27) has advocated over-exposed films to show the juxta-epiphyseal lesion to better advantage. It is sometimes advisable to obtain, in addition to conventional anteroposterior and lateral views of the hips, special projections, such as cone-studies of the affected side. In lateral projections, the best results in adults are obtained by using a small portable Bucky placed at the side to be examined. The central ray, paralleling the table top, passes under the normal thigh in the direction of the adductor surface of the affected leg. In children and infants, a telescopic cone can be used advantageously without the head-Bucky or Lysholm grid. When the hip is movable, it is more convenient to employ a flexed-thigh-abduction technic.

In previous communications (4, 66), we have called attention to the importance of the Morgan meter and Morgan-Hodges phototimer as a means of determining the proper technical factors for the examination in question. It is a rare technician who can approximate the excellence of this technic. Without the aid of the phototimer the following technics are employed.

Anteroposterior Technic: 100 ma.; kv.p., centimeter part thickness $\times 2 + 25$; 36 inches distance; Bucky grid; large or medium cone, depending on size of patient.

Lateral Technic: 50 ma.; 50-55 kv.p.; 28 inches distance; non-Bucky; 25° tilt toward the head; telescopic cone.

SUMMARY

1. We have attempted to present a fairly comprehensive review of the literature on the subject of the various types of dislocation, osteochondropathy, and congenital and adolescent coxa vara. The theories for the pathogenesis of these conditions have been briefly dealt with.

2. Illustrative case histories and roentgenograms from our service at the Newington Home and Hospital for Crippled Children have been presented, including rare lesions.

3. Attention has been directed to the importance of a thorough knowledge of normal anatomy, normal variants, and effective roentgen technics.

4. In a review of 300 normal hips of children up to six years of age our results have paralleled and corroborated those of Kleinberg and Lieberman.

5. The necessity for early diagnosis in congenital dislocation of the hip and Legg-Perthes disease has been emphasized in relation to the chances for a permanent cure.

NOTE: The authors desire to express sincere thanks for the help and interest of their colleagues of the Newington Home and Hospital staff. We are indebted to Dr. C. W. Hooker, Associate Professor of Anatomy of the Yale School of Medicine, for his assistance with the dissections, and to Mr. B. E. Foss for help with the photography.

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(For Spanish Summary, see following page)

SUMARIO

Lesiones de la Cadera en Lactantes y Niños Observados en el Asilo y Hospital Newington para Lisiados

Repásanse la embriología y la anatomía de la articulación coxal, con atención particular a las características radiográficas de la cadera normal. Recálcase la importancia del índice acetabular como auxiliar para evaluar la cavidad cotiloidea en los niños. Las mediciones de 300 caderas normales revelaron que el mismo es relativamente constante y concuerda con las observaciones de Kleinberg y Lieberman. En los lactantes de nueve días o menos el aclive medio resultó ser de 27° (de 10 a 44°). A la edad de doce meses había disminuído a 19 ó 20° , y a los cinco años a 11 ó 12° . En los niños pequeños la configuración de la cavidad puede simular una displasia acetabular en los mayores, y hay que mostrar cuidado para evitar un diagnóstico erróneo de displasia.

A base de sus observaciones en un asilo y hospital para niños lisiados, los AA. clasifican las enfermedades de la cadera en los lactantes y niños pequeños en la forma siguiente:

- I. Dislocación
 - A. Congénita
 - 1. Dislocación congénita
 - 2. Artrogriposis
 - 3. Dislocación asociada o secundaria a espina bífida

- B. Adquirida
 - 1. Traumática
 - 2. Séptica
 - 3. Enfermedad de Still
 - 4. Poliomieltis
 - 5. Subluxación en la distrofia muscular

II. Osteocondropatía (Enfermedad de Legg-Perthes)

III. Coxa vara y epifisiolisis congénitas

IV. Lesiones raras

- A. Miositis osificante
- B. Acondroplasia
- C. Osteogénesis imperfecta

Cada uno de dichos estados es discutido, considerándose también las teorías de la patogenia y presentándose típicas historias clínicas.

Hácese hincapié en la necesidad del diagnóstico temprano en la dislocación congénita de la cadera y en la enfermedad de Legg-Perthes en relación con las probabilidades de obtener la curación permanente. Una técnica roentgenológica exacta con las películas debidamente centradas y expuestas simétricamente, de preferencia con el enfermo erecto, es de la mayor importancia. A veces conviene obtener, además de las habituales vistas antero-posteriores y laterales de la cadera, proyecciones especiales, tales como estudios cónicos del lado afectado.

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The Radiologic Features of Neurofibromatosis¹

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Ann Arbor, Mich.

NEUROFIBROMATOSIS (von Recklinghausen) is a curious, often familial disease which in its severe forms is readily recognized by the association of multiple tumors of the peripheral nerves with areas of pigmentation in the skin, so-called *café au lait* spots. Other features, such as involvement of the central nervous system and the skeleton, are less frequently encountered but are of relatively greater importance to the radiologist. In order to investigate particularly the bone manifestations of this disease, we have reviewed the records and roentgenograms of 127 patients with von Recklinghausen's disease seen at the University of Michigan Hospital in the thirteen-year period between 1934 and 1947. Material derived from this study forms the basis for the present report.

HISTORICAL

Smith in 1849 comprehensively described neurofibromatosis, but failed to recognize the nerve-sheath origin of these multiple tumors (1). In spite of frequent additional references to molluscum fibrosum in the interim, it was not until 1882 that von Recklinghausen (2) established the true nature of the tumors and indelibly associated his name with this disease. He recognized that it was not an acquired condition and Thomson (3) in 1900 pointed out clearly its hereditary nature. Later, Preiser and Davenport (4) were able to show that neurofibromatosis appears in both sexes and follows the Mendelian law as a dominant trait.

In regard to bone changes, Adrian (5) reviewed the literature in 1901 and found scoliosis, skeletal anomalies, atrophy, and hypertrophy of individual bones and other defects frequently mentioned in descrip-

tions of patients with neurofibromatosis. Illustrations from Treve's account of the "elephant man" in 1885 show a clear-cut example of neurofibromatosis with bone involvement (6). Gould (7), in 1918, and Weiss (8), in 1921, commented on the frequent association of scoliosis. In 1922, Stahnke (9) recognized the varied skeletal lesions as an integral part of this disease and observed that many of the defects in bone apparently resulted from pressure, being closely associated with overlying soft-tissue neurofibromas. Brooks and Lehman (10) were the first to present a comprehensive classification of the skeletal changes in neurofibromatosis in the American literature. In 1924, they reported findings in seven patients and described three separate types of bone abnormality, including scoliosis, abnormalities of bone growth, and irregularity of outline of the shafts of long bones, "including changes which in the x-ray appear as subperiosteal cysts." Incomplete forms of von Recklinghausen's disease, including those with pigmentation only, had been described by Weber (6) in 1909 and, following this line of reasoning, Leader and Grand (11), in 1932, were able to tabulate 21 instances in which pigmentation in early childhood was followed in later years by typical neurofibromatosis. Grenet, Ducroquet, Isaac-Georges, and Macé (12) in 1934 described several examples of a *forme fruste*, with pigmentary and skeletal abnormalities. In one of the patients typical neurofibromatosis developed and, as the subjects of this report were children, the authors recognized the possibility that multiple neurofibromas might eventually appear in all. These same authors and later Ducroquet (13), in 1937, and Barber (14), in 1939, described a number of patients with neuro-

¹ From the Department of Roentgenology, University of Michigan, Ann Arbor, Mich. Presented at the Thirty-third Annual Meeting of the Radiological Society of North America, Boston, Mass., Nov. 30-Dec. 5, 1947.

fibromatosis who had bowing of a lower leg and pseudarthrosis following fracture or surgical attempts to correct the deformity. Regarding pseudarthrosis, Adrian's brief note that Fremmert (15) had described such abnormality following fracture in a patient with mollusum fibrosum in 1872 is of particular interest.

Uhlmann and Grossman (16), in 1940, reviewed the literature on neurofibromatosis and stated that 7 per cent of these patients showed osseous involvement varying from slight irregularity of bone contour to cyst-like cavities due to tumors embedded in bone. In the same year, Heublein, Pendergrass, and Widmann (17) discussed the relationship of neurofibromatosis to other neurocutaneous syndromes, *e.g.*, tuberous sclerosis, hemangiomas of the cerebrum with facial nevus, and von Hippel-Lindau's disease.

Many additional articles on the subject of neurofibromatosis have been written, but relatively few have appeared in the radiologic literature. Friedman's (18) account of osseous manifestations in neurofibromatosis and Doub's (19) editorial constitute the only articles we have found devoted exclusively to the subject.

INCIDENCE

The 127 cases of neurofibromatosis reported here were encountered in a period in which 245,219 new admissions were recorded at this institution. It is probable that the true incidence in the hospital population is somewhat higher, since many patients were not included either because of the incomplete nature of the physical findings or because the observations left some doubt in our minds that the condition encountered was actually von Recklinghausen's disease. A combination of the typical cutaneous and subcutaneous lesions with or without bone involvement was considered essential to the clinical diagnosis in patients included in this series. Sixty-three per cent had histologic proof. Although 130 cases of solitary neurofibroma seen in the same period were reviewed in the hope of uncovering additional examples

of von Recklinghausen's disease, this material has not been included in the present discussion. While it is by no means clear that such a distinction should be made, some authors believe that the individual tumors of multiple neurofibromatosis (von Recklinghausen) can be distinguished histologically from the solitary neurofibroma (20).

We found no important difference in incidence between the sexes, either with or without bone involvement, and observed no significant racial distribution. The age of patients varied from six weeks (condition present at birth) to seventy-seven years, although many patients gave a history of exacerbation or first appearance of tumors at about the time of puberty. The not infrequent familial incidence of this disease was manifested in nine individuals. We found no instances of occurrence or exacerbation of the disease during pregnancy, as has been reported (21).

PATHOLOGY

It has been suggested that neurofibromatosis be regarded as a disease which involves all of the sheaths and enveloping membranes of the nervous system (22). The individual nerve sheath tumor with its characteristic palisading of nuclei and streaming of cell bundles has been variously described as a neurilemoma, neurinoma, schwannoma, and perineurial fibroblastoma. It is beyond the scope of this paper to enter into the controversy which still exists as to whether these tumors arise from the mesodermal elements of the sheaths of nerves (20) or are derived from cells of neuroectodermal origin of the sheath of Schwann, as originally proposed by Verocay (23) and more recently emphasized by Masson (24).

Skin Manifestations: Using a special staining technic, Masson (25) was able to show that all pigmented nevi are true "nerve ending neuromas" developing from the peripheral sensory apparatus, a view which has gained general acceptance by pathologists. The *café au lait* spots in neurofibromatosis and the ordinary pig-

mented nevus, several of which can be found on the body of the average person, are indistinguishable histologically. In neurofibromatosis the pigmented areas tend to be large and distributed over the areas of most marked involvement of the nervous system or skeleton.

Soft pedunculated tumors which appear to have arisen from the skin (mollusum fibrosum) constitute another important cutaneous manifestation of von Recklinghausen's disease. They are of occasional interest to the radiologist because of the soft-tissue shadows which they may cast on films made for some other purpose.

Peripheral Nerve Lesions: The tumors associated with peripheral nerves may be discrete neurofibromas, in which case nerve fibers are said to enter and traverse the tumors (20). In other instances the peripheral nerve lesions are more diffuse in character, giving rise to the picture of so-called elephantiasis neuromatosa. The latter type of growth is characterized histologically as cirroid or plexiform neurofibroma. We have been impressed with the frequency with which osseous hypertrophy, and particularly periosteal new bone formation, occur in conjunction with this type of nerve lesion.

Central Nervous System Involvement: Twelve patients, or approximately 10 per cent, of our group showed convincing evidence of intracranial tumor, and in 8 cases proof was obtained by histologic or ventriculographic methods. These included meningiomas, astrocytomas, spongioblastomas, acoustic neuromas, and undifferentiated gliomas. In several patients the intracranial tumors were multiple. Other reports of frequently associated brain tumors have given the basis for considering von Recklinghausen's disease a defect in the supporting or "binding" tissues of the nervous system. Turner and Gardner (22) suggest that this concept be extended to include the glial elements as well.

The frequent association of neurofibromatosis with acoustic neuromas has been generally recognized, and Gardner and Frazier (26) report a series in which bi-



Fig. 1. Case No. 301303. Typical saucer-shaped erosions on posterior aspect of femur due to adjacent neurofibromas. These are the so-called "pits" or "caves" described by Nørgaard as the most characteristic sign of neurofibromatous bone involvement.

lateral tumors of the acoustic nerves were associated with neurofibromatosis in 84 per cent of the cases. In a survey of 160 cases of acoustic neuroma, Gonzalez-Revilla (27) found only 6 examples of bilateral involvement. Three of these patients had generalized neurofibromatosis.

Intradural neurofibromas occasionally appear to arise from the spinal cord, but tumors originating in sheaths of spinal nerve roots are much more common. As such they may be confined to the spinal canal or may appear as intrathoracic or intra-abdominal tumors. Some of these lesions may present a "dumbbell" shape, projecting to either side of the spinal nerve root canal, producing pedicular erosion and widening of intervertebral foramina at the waist of the tumor.

Malignancy: Malignant degeneration occurring in a neurofibroma is not infre-

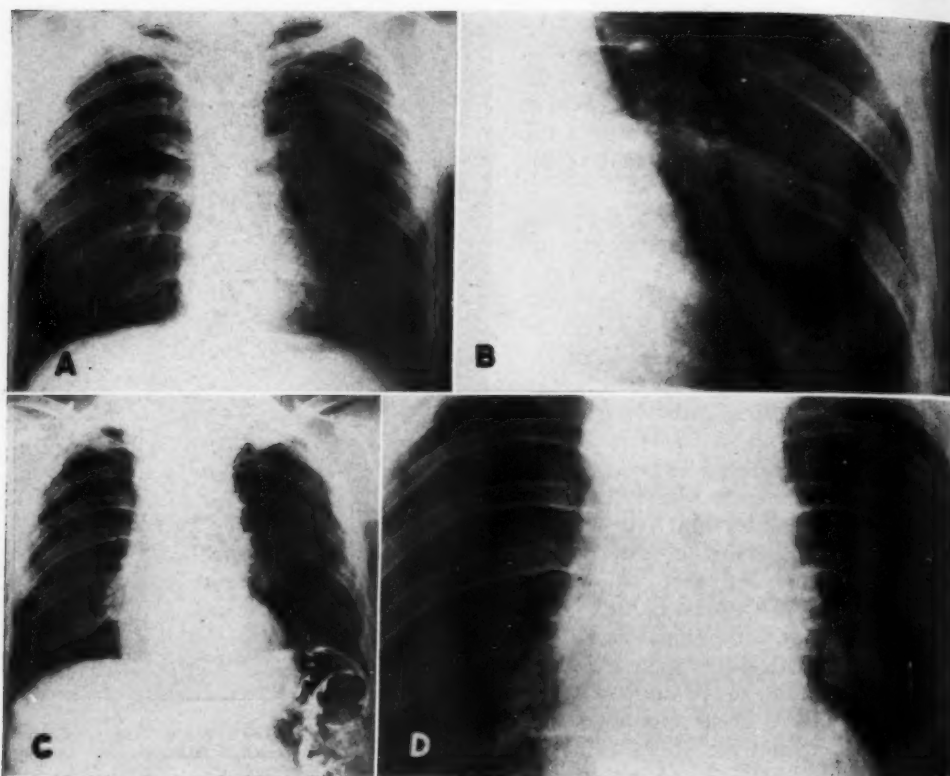


Fig. 2. A and B. Case No. 368691. Chest film of 25-year-old man who has multiple neurofibromas distributed along nerve trunks of entire body. Tumors of intercostal nerves have produced irregularity and erosion of inferior surfaces of several ribs simulating the supposedly pathognomonic notching of aortic coarctation.
C and D. Case No. 599723. More pronounced scalloping of ribs in 18-year-old male with typical von Recklinghausen's disease. Mediastinal and intercostal tumors more clearly evident than in preceding case.

quent. It was reported in 12 per cent of cases reviewed by Hosoi (28). In his review of 65 case reports, the age ranged from fifteen to seventy years and while prognosis was, of course, poor, metastases occurred in only 22 per cent. Miller (29) reported a case in which there was simultaneous malignant degeneration in several neurofibromas. In our series there were 7 instances of malignant change in neurofibroma, an incidence of 5.5 per cent. The lesions were reported by the pathologist as either neurofibrosarcoma, fibrosarcoma, or spindle-cell sarcoma.

BONE INVOLVEMENT

The primary purpose of our study was to investigate the osseous manifestations of

neurofibromatosis, and we have been amazed at the frequency, variety, yet consistent pattern encountered in these osseous lesions. Thirty-seven patients, or slightly over 29 per cent of the total group, showed some form of skeletal defect which could be correlated with the clinical manifestations of the disease. It should be understood that no attempt is being made to present this figure as an accurate index of bone involvement in neurofibromatosis, since in 50 of our 127 cases there was no investigation of the skeletal system by roentgenographic means. Furthermore, many persons with milder forms of von Recklinghausen's disease undoubtedly never present themselves for medical treatment of their seemingly insignificant disorder.

While at first glance the bone changes seem to show endless variety, review of a large number of cases establishes a few distinct types of skeletal abnormality under which the majority may be classified. These types may be listed as follows:

1. Erosive defects due to the presence of neurofibromas contiguous to bone.
2. Scoliosis.
3. Disorders of growth, including both over- and under-development.
4. Bowing and pseudarthrosis of the lower leg.
5. Intra-osseous cystic lesions.
6. Congenital anomalies.

Of these, scoliosis and congenital anomalies seem to be the most prevalent, but bowing and pseudarthrosis, localized overgrowth or atrophy, and defects in bone contiguous to neurofibroma are the most characteristic.

1. *Erosive Changes:* Nørgaard (30) states that the characteristic bone change associated with neurofibromatosis is the "pit or cave" due to erosion of the bone by an adjacent neurofibroma (Fig. 1). While such defects are characteristically observed in the bones of the extremities, we have encountered two cases in which neurofibromas of intercostal nerves produced bilaterally symmetrical notching of several ribs with a superficial resemblance to the supposedly pathognomonic sign of coarctation of the aorta (Fig. 2). In each instance soft-tissue tumors in close proximity to the "notching" of the ribs were noticed on closer inspection and, together with incontrovertible clinical evidence of neurofibromatosis, led to the proper evaluation of these defects. Although these rib changes differed in no way from the pressure defects described by Stahnke (9), the pits or caves of Nørgaard (30), or the irregularity of outline of shafts of long bones noted by Brooks and Lehman (10), specific involvement of ribs has been mentioned rarely, and the similarity of such defects to those seen in coarctation of the aorta has not been described previously to our knowledge.



Fig. 3. Case No. 536736. Fourteen-year-old boy with *café au lait* spots and multiple subcutaneous tumors. Extensive erosion of left side of upper thoracic spine and adjoining ribs due to neurofibroma arising within the spinal canal and extending into the thorax. Note the twisted-ribbon appearance of the fourth and fifth ribs. An intrathoracic neurofibroma was resected.

Localized erosion of the posterior portions of the ribs in the upper thorax is usually encountered in conjunction with similar defects in corresponding vertebrae (16, 29) (Fig. 3). There probably is nothing precisely characteristic about this type of rib erosion, but as it is so infrequently associated with intrathoracic tumors other than neurofibromata, it must be regarded as having considerable diagnostic significance.

2. *Scoliosis:* Erosive changes in the spine usually occur asymmetrically, producing structural defects which result in abnormal curvature. Although erosion may occur in any portion of the vertebral column, Miller (29) states that the typical scoliotic deformity occurs in the lower dorsal region and consists of a kyphoscoliosis in which the kyphotic element predominates. Several patients in our group have shown cervical kyphosis of varying degrees of severity, as illustrated in Figure 4.

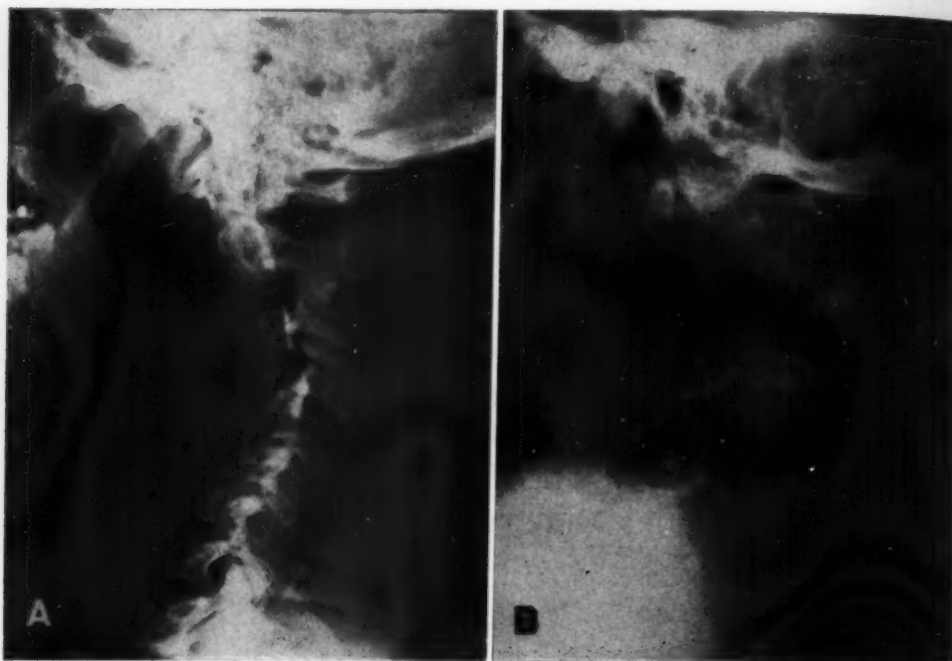


Fig. 4. A. Case No. 401873. This 23-year-old woman with neurofibromatosis had a thoracotomy which showed "extensive plexiform neurofibroma of lower cervical and upper thoracic nerve roots." The moderate cervical kyphosis probably is due to bone erosion by adjacent mid-cervical neurofibroma. See Fig. 7.

B. Case No. 586909. Similar but more pronounced deformity in 24-year-old woman with skin pigmentation, subcutaneous tumors and bizarre anomalies of hands and feet. Progressive transverse myelitis for period of one year at level of C-3.

In some patients with neurofibromatosis, scoliosis of varying degree is readily explainable on the basis of over-growth or under-growth of a lower extremity. In other patients with the disease, there may be coexisting anomalous development of one or more vertebrae which will result in eventual abnormal curvature. Whatever the cause, it is clearly evident that scoliosis is the commonest osseous defect occurring in conjunction with neurofibromatosis.

3. *Disorders of Growth:* Among disorders of growth, the most frequently encountered in our series of cases was overgrowth of one or more bones. It is significant that in all such instances the elephantiasic soft tissues adjacent to the bones involved were infiltrated by cirroid or plexiform neurofibroma. Histologic examination of tissue removed from the lower leg of one patient showed cirroid neurofibroma invading and elevating the

periosteum; subsequent roentgenograms demonstrated slow but progressive subperiosteal new bone formation over a ten-year period. These pathological and roentgen findings adequately explain the increased width of the bones involved, but scarcely account for the acceleration of longitudinal growth which also occurred in this patient (Fig. 5). Furthermore, similar hypertrophy was found in various bones of six other individuals in whom periosteal thickening was either a less conspicuous feature or was altogether absent. In addition to osseous hypertrophy, accelerated epiphyseal ossification was observed occasionally.

The exact mechanism by which longitudinal overgrowth is produced in neurofibromatosis is not clearly understood, but increased blood supply resulting from lymph stasis offers the most plausible explanation. Incidentally, pronounced over-

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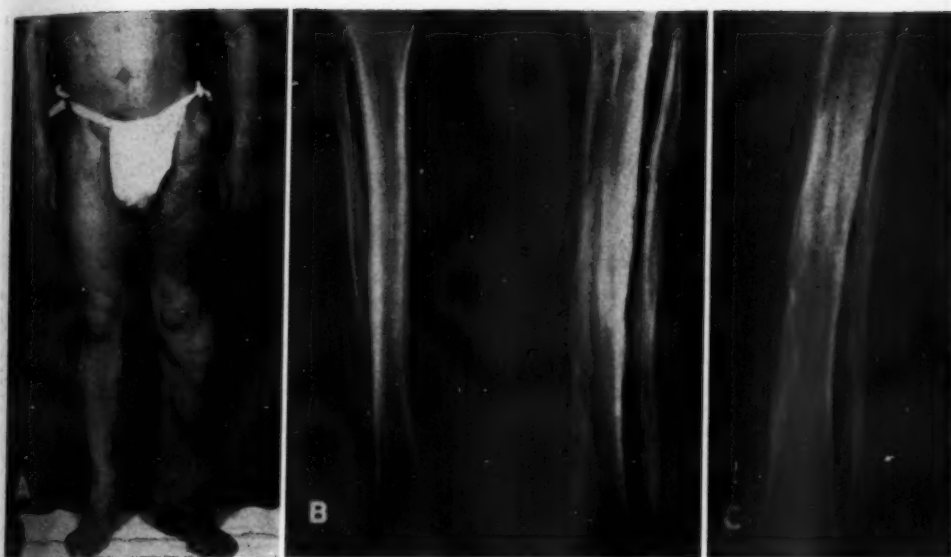


Fig. 5. Case No. 301303. A. Thirty-three-year-old man with neurofibromatous elephantiasis of left leg. Areas of skin pigmentation are clearly visible on both thighs, chest, and abdomen. The left leg is five inches longer than the right.

B. Longitudinal overgrowth of left tibia and fibula and a fairly characteristic type of periosteal proliferation similar to that occurring in patients reported by Brooks and Lehman (10) and Friedman (18).

C. Left leg 10 years later. There is a small periosteal "cyst" or "bleb" in the distal portion of the fibula (arrow). For roentgenographic appearance of distal left femur, see Fig. 1. Left leg amputated (mid thigh). Specimen showed "plexiform neurofibroma infiltrating and replacing periosteum" but not involving cortex or medullary cavity.

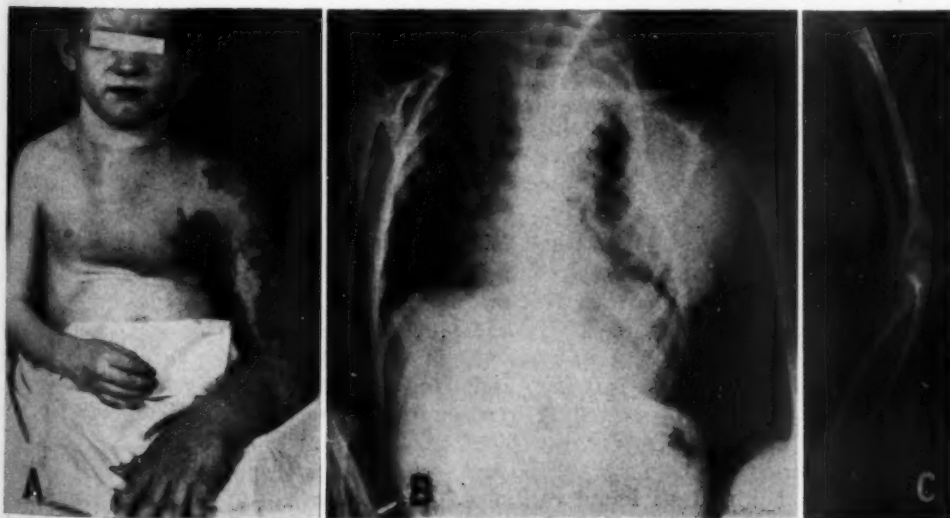


Fig. 6. Case No. 439090. A. Hyperpigmentation, overgrowth, and accelerated maturation of left arm and hand of 4-year-old boy. Extension of plexiform neurofibroma (biopsy) into thorax and left side of neck. Lesion, present since birth, was treated by x-ray therapy without response.

B. Deformity of thoracic cage and beginning scoliosis presumably due to excess weight of huge left arm.

C. Marked overconstriction of left humerus, radius, and ulna. Maintenance of normal bone density suggests trophic change rather than pressure atrophy.

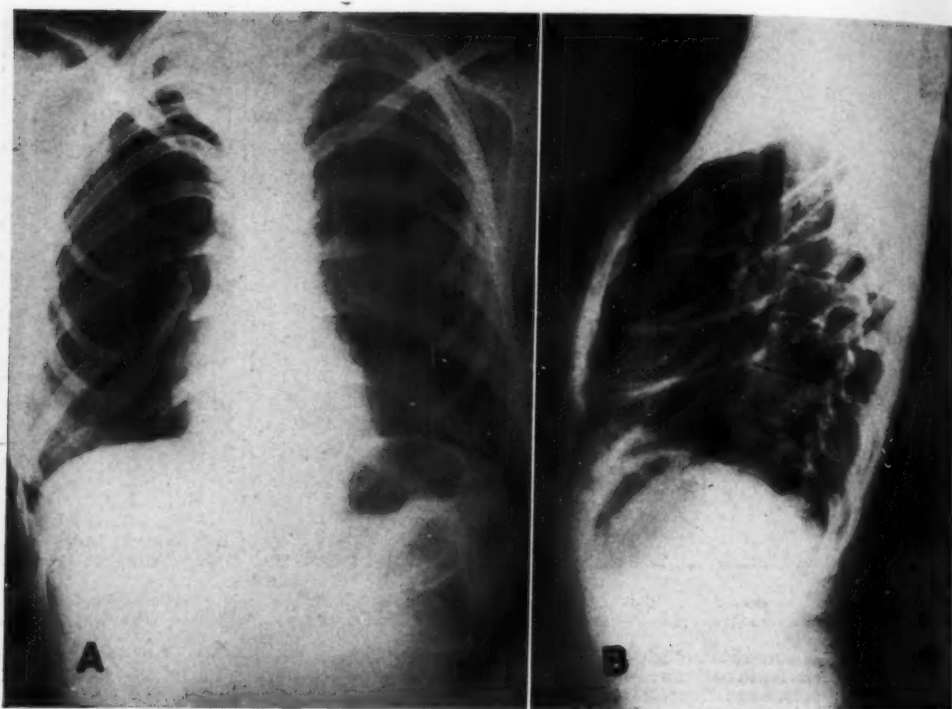


Fig. 7. Case No. 401873. Twenty-three-year-old woman whose cervical spine is shown in Fig. 4 A. Overly constricted ribs correspond segmentally to distribution of upper thoracic neurofibroma. Anterior and axillary portions of ribs are more markedly narrowed than posterior segments which are in direct contact with the soft-tissue tumor. Rib defects such as these occurring in conjunction with an intrathoracic tumor suggest neurofibroma as most likely tumor type.

growth of soft tissues in so-called *elephantiasis neuromatosa* can be identified roentgenographically and used as a helpful point in differential diagnosis (Figs. 5 and 6).

Although hypertrophy of a long bone in neurofibromatosis usually is circumferential as well as longitudinal (16), two patients in our group showed actual diminution in the caliber of bones which were unquestionably increased in length. In one patient, a child of four years with a diffuse cirroid neurofibroma of the left arm and hand, all the bones of this extremity were involved in such a manner (Fig. 6). The apparent over-constriction of the bones was quite similar to that found in longstanding paralytic conditions, such as old poliomyelitis, although one notable difference was evident; in neurofibromatosis the calcium content of the atrophic bones appeared to be entirely normal.

The second patient had involvement of the left foot, and the metatarsals were similarly elongated and narrowed. It is of further interest that this patient eventually required amputation of the left foot (mid leg), and postoperatively the fibula underwent progressive shrivelling until it almost completely disappeared; this despite the fact that amputation was done high above the site of soft-tissue neurofibroma. In both the slender metatarsals and the subsequently narrowed fibula, approximately normal bone density was maintained.

Similar narrowing and sinuosity of bone with maintenance of normal calcium content were observed in the ribs of at least two other patients with neurofibromatosis, one of whom had moderate intrathoracic extension of a tumor arising within the spine (Fig. 7). In this particular individual

the anterior portions of the ribs were more profoundly narrowed than the posterior portions immediately adjacent to the spinal neurofibroma, suggesting a growth defect occurring early in embryogenesis. That such an explanation is not adequate in all such cases is exemplified by the previously mentioned individual whose fibula withered spontaneously following amputation, and by still another patient whose right clavicle was seen to have become markedly narrowed and tortuous eleven years following successful operative removal of a supraclavicular neurofibroma.

The methods by which such change occurs in neurofibromatosis is a matter for conjecture. Pressure atrophy *per se* cannot be accepted as the prime factor because in some of the cases no tumor is to be found immediately adjacent to the bone involved. Furthermore, the osteoporosis which is so characteristic of pressure phenomena in bone is notably absent. One might better consider these changes as being of the so-called "trophic" variety, providing he is not implying the presence of highly mythical "trophic nerves," but rather is accepting the modern conception of trophesis, namely that it represents a disturbance of nutrition incident to faulty innervation, inadequate blood supply, absence of work-producing stimuli, etc.

It has been reported by Brooks and Lehman (10) that shortening of a growing bone will result if neurofibromatous tissue invades the epiphysis. We encountered no authenticated instances of such osseous undergrowth in our series of cases.

4. *Congenital Bowing and Pseudarthrosis of the Lower Leg:* The frequent association of neurofibromatosis and congenital bowing of the tibia (with subsequently developing pseudarthrosis) is generally accepted. Yet several recent reports (31, 32) describe the tibial deformity without the typical cutaneous and subcutaneous neurofibromas.

Can congenital bowing of the tibia be considered an integral part of von Recklinghausen's disease? Those who support such a theory point out that multiple

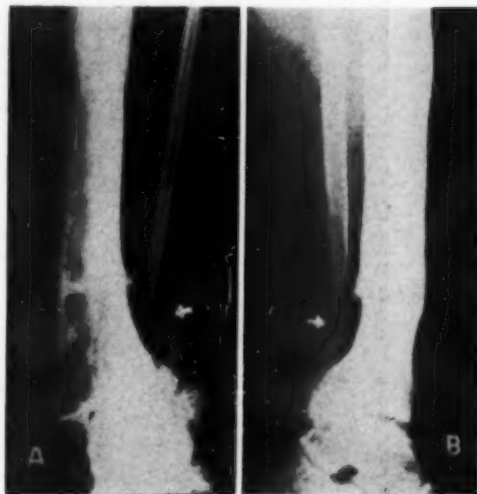


Fig. 8. Case No. 155132. Seventeen-year-old girl whose lower leg was deformed since birth. No pigmentation of skin and no soft-tissue tumors in childhood. Pseudarthrosis of left tibia and fibula followed fracture which resulted from trivial injury at age of eight years. Tibia eventually healed with aid of bone graft but fibular pseudarthrosis persisted. A small soft-tissue tumor which formed adjacent to the fibular pseudarthrosis when the patient was twenty years old proved histologically to be "a neurofibroma."

neurofibromas first appear at puberty or later in many cases, while the tibial bowing is present at birth. The situation is perhaps analogous to the finding of large *café au lait* spots in a child without other evidence of neurofibromatosis. Ducroquet and his co-workers (12, 13) consider the pigmentary and osseous abnormalities in such cases to be either an incomplete form (*forme fruste*) of von Recklinghausen's disease or the precursor of the more characteristic cutaneous and subcutaneous tumors. The sequence of events in one case reviewed by us seems to add support to this somewhat extreme view. An eight-year-old girl had pseudarthrosis of the bowed left tibia and fibula developing after a trivial injury. The tibia eventually united with graft but the fibula did not. The patient was again seen at twenty years of age, when a nodule removed from the soft tissues near the fibular pseudarthrosis was found to be a neurofibroma (Fig. 8).

The present state of our knowledge does



Fig. 9. Case No. 558528. A and B. Jan. 23, 1945. Congenital anterolateral bowing of the left tibia and, to a lesser extent, of the left fibula. There is an elongated area of rarefaction at the junction of the upper and middle thirds of the tibia. Trabecular pattern of mid portion of tibia shows considerable distortion.

C and D. Dec. 27, 1946. Persistent pseudarthrosis following fractures of left tibia and fibula. Amputation eventually deemed advisable. Multiple histologic sections of both bone and adjacent soft tissues showed no neurofibromatous tissue.

not seem to justify the outright conclusion that congenital bowing and pseudarthrosis of a lower leg is a pathognomonic sign of von Recklinghausen's disease. Long-range observation of a large number of these patients and their families would be necessary to substantiate such an opinion.

The mechanism by which the bowing and pseudarthrosis occur in those patients with neurofibromatosis has received much attention but no completely satisfactory explanation. Moore (33) believes the nerve pathology and the pseudarthrosis "stand in the relation of cause and effect." Green and Rudo (34) report the presence of intraosseous neurofibromatous tissue at the site of deformity, a finding which, if present in all instances of congenital bowing, would readily explain the occurrence of fractures and their inability to heal. An

excellent opportunity for the investigation of this possibility was afforded us when one patient with long-standing pseudarthrosis required amputation (Fig. 9). Extensive histologic search failed to reveal neurofibromatous tissue in or near the site of deformity. The additional possibility must be considered that the bony deformity is an associated anomaly of the skeleton occurring in a disease which itself appears to be a dystrophy primarily of neuroectodermal origin, but with participation of mesodermal elements.

We encountered six patients with congenital bowing and some clinical evidence of neurofibromatosis; all but one developed pseudarthrosis at the fracture site or in an osteotomy which had been done in an attempt to correct the deformity. The exception is a twenty-one-month-old

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Fig. 10. A. Oct. 20, 1936. This patient, a 4-month-old girl, had multiple subcutaneous tumors proved by biopsy to be neurofibromas. A routine bone survey showed the above intra-osseous "cystic" lesions and peripheral saucer-shaped erosions in the metaphyses of the femurs, tibiae, and fibulae. The upper extremities were involved to a lesser degree. The Kahn reaction was negative.

B. Nov. 16, 1937. Complete spontaneous disappearance of osseous lesions as well as soft-tissue tumors. Child has remained well. (Courtesy Dr. M. J. Cooperstock, Marquette, Mich.)

boy who has been followed for less than six months, during which time the extremity has been protected with the hope of avoiding fracture. In two patients in this group solid bony union eventually developed following repeated osteotomy; one required amputation, and two patients showed no union when last seen at this institution. These patients present a difficult orthopedic problem until they approach the age of puberty when, for some reason, the chance of successful bone graft fixation of fracture increases.

5. *Intra-osseous Cystic Lesions:* The occurrence of cystic lesions is perhaps the most interesting and possibly the most controversial aspect of bone involvement in neurofibromatosis. Brooks and Lehman (10) described "subperiosteal cysts" and postulated an interesting mechanism by which these changes may occur. They suggested that a neurofibroma arising in a

periosteal nerve erodes the adjacent bone and comes to lie partially or entirely within it. Proliferation of the osteogenetic elements in the periosteum produces a covering of bone over the neurofibroma. Various stages of this process were observed in one of our patients (Fig. 5) and, whereas the diffuse neurofibromatous tissue probably originated in overlying soft tissues rather than bone, microscopic sections showed invasion and elevation of the periosteum.

De Santo and Burgess (35) present evidence for an intrinsic nerve supply for cortical, cancellous, and medullary bone as well as periosteum, and several authors (18, 35, 36, 37) report histologically proved examples of intra-osseous neurofibromas. Some authorities, on the other hand, feel that the presence of nerves within bone is unproved and, therefore, doubt the actual intra-osseous origin of such lesions.

Apart from the subperiosteal cyst, which

seems to be an established entity in this disease, we have observed a number of cystic lesions within cortical and cancellous bone in patients with neurofibromatosis. In several individuals with classical von Recklinghausen's disease, a bone survey demonstrated localized areas of rarefaction sharply outlined by thin shells of marginal density. In two instances these lesions were in the distal femoral metaphyses and suggested the areas of localized fibrous dysplasia not infrequently seen in growing children. In neither case was a bone biopsy justified in view of other findings. These lesions were similar to that reported by Leader and Grand (11), who also presented no histologic proof that the osseous defect actually contained neurofibromatous tissue. Khoo (38) suggests "that the finding of one or several isolated bone cysts is as suggestive of the existence of Recklinghausen's neurofibromatosis as the presence of an isolated neurofibroma of the skin or of one or more *café au lait* spots."

Two other patients showing multiple intra-osseous cystic lesions are probably the most remarkable in our entire series. The first infant was seen at the age of four months and a clinical and histologic diagnosis of neurofibromatosis was established. At this time a skeletal survey disclosed numerous intra-osseous cystic lesions lying within the metaphyses of many of the long bones (Fig. 10A). Thirteen months later, at which time the subcutaneous lesions had regressed considerably, the osseous changes had completely disappeared (Fig. 10B). To our knowledge spontaneous regression of multiple bone lesions in neurofibromatosis has not been previously described.

The second child has been studied more recently. Multiple fixed and movable tumors were noticed over the trunk and extremities a few hours after birth. At this hospital, at the age of six weeks, roentgenograms showed the startling involvement of the long bones, spine, and ribs shown in Figure 11. Only the skull was spared. In the long bones the distribution and configuration of lesions were quite

similar to those seen in the preceding case, and on the basis of our previous experience a roentgen diagnosis of neurofibromatosis was suggested. A biopsy of the soft tissues demonstrated neurofibroma with calcification in a necrotic center, but three biopsies of the intra-osseous lesions were unsuccessful in establishing the presence of neurofibromatous tissue within bone. This child has not been followed long enough to determine whether or not the lesions will show the striking regression observed in the earlier case.²

It is this group of cases that has forced us to keep an open mind as to the possible relationship of neurofibromatosis to so-called fibrous dysplasia as well as certain other abnormalities of bone. Thannhauser (39) has suggested a common etiology for neurofibromatosis and fibrous dysplasia but Jaffe (40) has denied any such link.

6. *Associated Anomalies of the Skeleton:* There is every reason to expect a high percentage of congenital anomalies in this disease, which itself seems to result from some defect in the chromosomes and genes passed on to the individual by his forebears. In fact, some of the bone changes which have been described may be nothing more than associated anomalies, e.g., congenital bowing of the tibia. Although spina bifida, fusion of vertebral bodies, congenital dislocation of the hips, clubfoot, and other anomalies have been frequently described in von Recklinghausen's disease, we have not been particularly impressed by the high incidence of such findings in our series and have made no attempt to tabulate the incidence of skeletal anomalies. In one patient, symmetrical anomalies of the hands and feet were so striking as to deserve special mention.

LeWald (41) described four patients with congenital absence of the superior orbital wall associated with pulsating ex-

² Recent examination of this patient at the age of eight months shows virtually complete spontaneous disappearance of the subcutaneous tumors mentioned above and remarkable regression of the osseous defects illustrated in Figure 11.



Fig. 11. Case No. 611263. A 6-week-old boy who had innumerable subcutaneous tumors, first noticed twelve hours after birth. Many of the tumors were freely movable; others were attached to bone. Biopsy of soft-tissue tumor showed proliferating neurofibroma with central area of necrosis and calcification. Kahn test negative. Serum calcium 9.2 mg. per cent; phosphorus 6.6 mg. per cent; alkaline phosphatase 13 Bodansky units.

A. Roentgenograms of lower extremities show saucer-like erosions of distal femurs and proximal tibias as well as metaphyseal intra-osseous cystic lesions similar to those shown in Fig. 10. Numerous areas of soft-tissue calcification are present.

B. Extensive areas of bone erosion and destruction are seen in the ribs, spine, ilia, and pectoral girdles. Scoliosis is due to destructive lesion in right half of L-1.

C. Right upper extremity shows metaphyseal rarefactions and subcutaneous soft tissue tumors (the left arm was similar in appearance). Destructive lesions also were observed in the mandible, but the cranial vault was entirely normal.

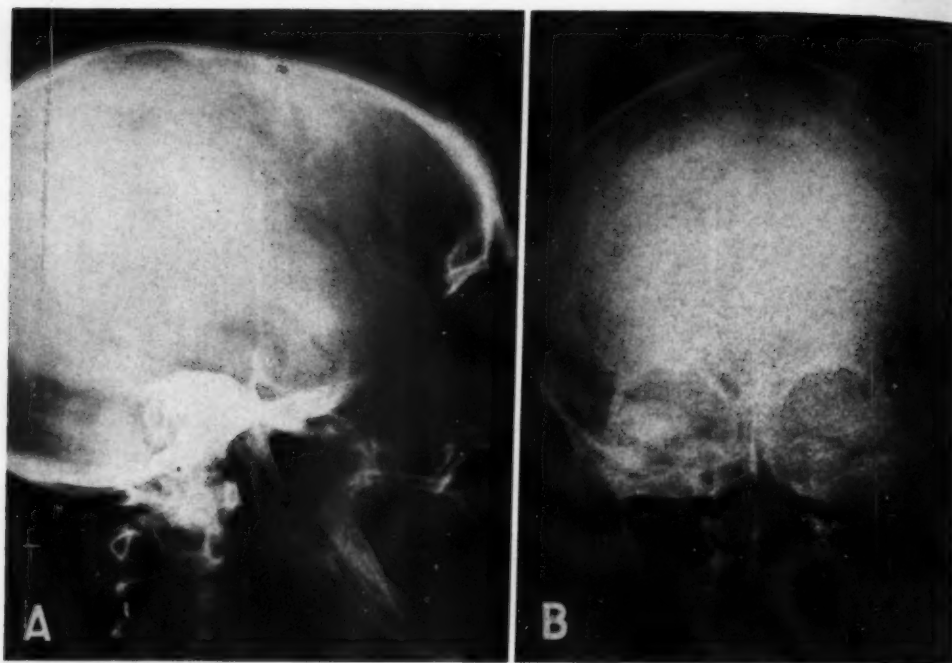


Fig. 12. Case No. 352063. Twenty-seven-year-old woman with generalized neurofibromatosis, initially seen at University Hospital in 1935, at which time the first of multiple plastic procedures was done for treatment of neurofibromas of eyelids. Skull films, requested in connection with this survey, show virtually complete absence of the sphenoid bone. There was no neurologic evidence of an intracranial tumor.

ophthalmos, and two of these had von Recklinghausen's disease. Reviewing the literature, Peyton and Simmons (42) were able to find twenty case reports of neurofibromatosis with orbital wall defects and added five cases of their own. Such patients generally have demonstrable neurofibromatous tissue within the eyelids or orbital soft tissues, and it is open to question whether the orbital wall defects occur as the result of erosion by the adjacent neurofibroma or represent a coexisting congenital defect. Pulsating exophthalmos is a prominent feature which is said to result from transmitted cerebral pulsation.

We were able to find two patients of this type in our group and in one the osseous abnormality had been unsuspected over a period of several years, during which plastic procedures were done for the disfiguring and disabling neurofibromatous involvement of the eyelids. In view of the known frequency of associated bone

changes, we requested roentgenograms of the skull and orbits and were rewarded by the astonishing findings seen in Figure 12. Not only were the walls of both orbits absent, but the floor of the anterior cranial fossa was virtually non-existent. It must be admitted that the pointed configuration of the dorsum sellae suggests erosion, but there was no neurologic evidence of intracranial tumor. Other than the mechanical interference produced by the abnormal lids, vision was well maintained. Incidentally, this patient's son also had neurofibromatosis, involving the tongue and lower extremities.

DIFFERENTIAL DIAGNOSIS

None of the bone changes associated with von Recklinghausen's disease can be considered pathognomonic, but they are frequently strongly suggestive of this condition. The differential diagnosis varies with the type of bone lesion encountered.

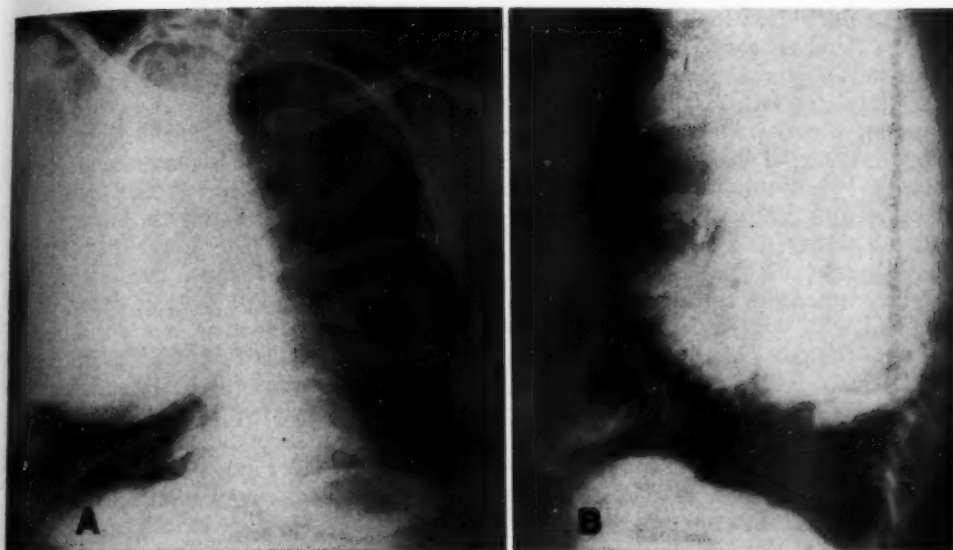


Fig. 13. Case No. 609852. Huge meningocele protruding through a 3×5 -cm. defect in the right lateral portion of the upper thoracic spine. Smooth configuration of tumor in conjunction with multiple rib defects, scoliosis, and *café au lait* spots led to erroneous preoperative diagnosis of intrathoracic neurofibroma.

Erosive changes may occur with other tumors, aneurysms, and, in respect to ribs, in coarctation of the aorta. Smooth erosion, particularly in characteristic locations such as the spine and adjacent ribs, prompts serious consideration of neurofibromatosis. One patient offered an interesting problem in diagnosis. She was a woman thirty-six years old, with a large, smoothly rounded tumor in the posterior portion of the right hemithorax. As there were associated erosion and deformity of vertebrae and ribs along with upper dorsal scoliosis, an unqualified roentgenologic diagnosis of neurofibromatosis was made. This opinion seemed to be thoroughly substantiated when it was discovered that the patient had multiple *café au lait* spots. At thoracotomy, however, the tumor was found to be cystic in nature, and a careful dissection revealed it to be a large antero-lateral meningocele, which was successfully resected. The case (43) will be reported in detail in another journal.

The causes of scoliosis are so numerous that the radiologist will seldom, if ever, suspect neurofibromatosis from this finding alone.

Overgrowth of individual bones occurs in hemangioma, lymphangioma, chronic osteomyelitis, and so-called hemihypertrophy, as well as in neurofibromatosis. Accelerated maturation and hypertrophy of epiphyses often occur in hemophilia as the result of recurrent hemarthrosis. Irregular periosteal thickening, which may be a part of the generalized bony overgrowth in neurofibromatosis, may suggest either specific or non-specific osteomyelitis (44). Atrophy or underdevelopment of bones is found in post-paralytic states and in various debilitating diseases. A helpful differential point is the absence of osteoporosis in the atrophy associated with neurofibromatosis. The thin, unusually gracile bones of osteogenesis imperfecta may resemble the atrophic bones in neurofibromatosis, but in the former the changes are more apt to be generalized.

Bowing of the lower leg frequently occurs in osteogenesis imperfecta, and pseudarthrosis may follow one of the many fractures sustained by these patients. Ordinary fractures also may fail to unite, in which case they will present the picture of pseudarthrosis. Rickets may result in

bowing of the tibia, which usually will not have the anterior curve so characteristically found in neurofibromatosis.

The intra-osseous cystic lesions may suggest an almost infinite number of conditions. A partial list would include so-called fibrous dysplasia of the localized type as well as that seen in Albright's syndrome, bone cyst, hyperparathyroidism, Ollier's disease, and the reticulosos.

Skeletal anomalies are no different when associated with von Recklinghausen's disease, with the possible exception of orbital wall defects. Occasionally, these orbital changes may be imitated by other tumors, cysts, or aneurysms.

Whatever the type of bone abnormality, the radiologist will be wise to insist upon a complete evaluation of the patient before concluding that the osseous lesion results from neurofibromatosis. In the majority of instances there will be sound clinical evidence of von Recklinghausen's disease and the osseous manifestations will form an integral part of the syndrome. In a significant number of cases the cutaneous and subcutaneous lesions will be easily overlooked or misinterpreted, and the radiologist will have the opportunity to first suggest the diagnosis from the osseous lesions.

SUMMARY

Since von Recklinghausen's classical description of neurofibromatosis in 1882, numerous case reports have emphasized the frequency and variability of associated bone changes. It is quite evident that the disease is not a rarity, and equally apparent that roentgen methods are most useful in evaluating the osseous changes which occur in conjunction with it.

After reviewing 127 case reports of patients with neurofibromatosis it seems to us that the incidence of bone involvement is considerably greater than the figure of 7 per cent commonly quoted in the literature. Furthermore, because of the wide variety of bone changes which may occur, we believe that neurofibromatosis should be considered more frequently in the dif-

ferential diagnosis of obscure skeletal abnormalities. Likewise, roentgenologic skeletal surveys of patients with known von Recklinghausen's disease are apt to show entirely unsuspected osseous defects.

In general the bone lesions in neurofibromatosis may be classified as erosive defects, scoliosis, disorders of growth, bowing and pseudarthrosis of the lower leg, intra-osseous "cystic" lesions, and associated congenital anomalies. In this paper, each type of osseous lesion is considered separately, and various facts and theories are reviewed in regard to the mechanism by which each is produced.

Two interesting manifestations of neurofibromatosis deserving particular mention are spontaneous regression of multiple intra-osseous "cystic" lesions and erosive changes simulating the rib notching in aortic coarctation.

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SUMARIO

Características Radiológicas de la Neurofibromatosis

Desde la clásica descripción de la neurofibromatosis por von Recklinghausen en 1882, numerosas observaciones clínicas han recalado la frecuencia y variabilidad de la patología ósea concomitante. Es manifiesto que la enfermedad no constituye una rareza, e igualmente que las técnicas roentgenológicas son de la mayor utilidad para

justipreciar las alteraciones óseas que sobrevienen en la misma.

Después de repasar 127 historias clínicas de neurofibromatosis, les parece a los AA. que la incidencia de la invasión ósea es considerablemente mayor que el 7 por ciento citado habitualmente en la literatura. Además, debido a la gran variedad

de alteraciones que pueden ocurrir, opinan que debe considerarse más a menudo la neurofibromatosis en el diagnóstico diferencial de las anomalías oscuras del esqueleto. Así también, los estudios radiológicos del esqueleto de los sujetos con enfermedad de von Recklinghausen reconocida son susceptibles de revelar deformidades óseas absolutamente insospechadas.

En general, cabe clasificar las lesiones óseas de la neurofibromatosis en deformidades erosivas, escoliosis, trastornos del desarrollo, encorvamiento y pseudoartrosis de la porción inferior de la pierna, lesiones "quísticas" intraóseas y anomalías congénitas asociadas. En este trabajo, considérase por separado cada forma de lesión

ósea y se analizan varios datos y teorías en relación con el mecanismo que produce cada una de ellas.

No puede aceptarse como patognomónica a ninguna de las alteraciones asociadas con la enfermedad, pero frecuentemente resultan muy indicativas. El diagnóstico diferencial varía conforme a la forma de la lesión ósea descubierta.

Dos interesantes manifestaciones de la neurofibromatosis que merecen cuidadosa atención son la regresión espontánea de las lesiones "quísticas" intraóseas múltiples y las alteraciones erosivas que simulan la escotadura costal en la estenosis de la aorta.



Sickle-Cell Anemia in Adults: Roentgenographic Findings¹

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New York, N. Y.

SICKLE-CELL anemia is a chronic hemolytic anemia occurring almost solely in the Negro race. It is characterized by dyspnea, fleeting pains in the extremities, leg ulcers, attacks of severe abdominal pain, anemia, and the presence of sickle-shaped red blood cells.

The occurrence of "peculiar elongated and sickle-shaped red blood corpuscles in a case of severe anemia" was observed in a twenty-year-old Negro man by Herrick in 1904. Later he reported (15) the case "because of the unusual blood findings." Herrick said he did not know "whether the blood picture is dependent on some peculiar physical or chemical condition of the blood or is characteristic of some particular disease," but that "if a similar condition is found in some other case a comparison of clinical conditions may help in solving the problem." Herrick's observations were confirmed by Washburn (37), Cook and Meyer (7), Mason (23), and Sydenstricker (32). A comparison of the clinical and blood findings showed the condition to be a disease entity previously undescribed.

An inherited abnormality in the stroma of the red blood cell is the underlying cause of the disease. The curious sickle-shaped cells are interpreted as being a manifestation of this defect (14). It is estimated that about 7 per cent of the Negro population show the phenomenon of sickling (sickleemia), while only about 1 to 2 per cent of those showing sickleemia develop a severe hemolytic anemia (sickle-cell anemia) (24, 32).

The mortality rate from sickle-cell anemia is not easy to determine. It is probable that there is great variation in susceptibility to the disease and in its severity. Many of those affected die dur-

ing infancy and childhood, although some complete a normal span of life (11, 24, 32). The terminal clinical picture in a large number of fatal cases is described as one of extreme prostration, peripheral vascular collapse, and shock (34). At necropsy the findings are mainly vascular lesions, infarction, thrombosis, and focal areas of necrosis. Extensive erythrophagocytosis is found in the spleen, liver, lymph nodes, and bone marrow (1, 11, 31, 32). It has been suggested that the sickle-shaped cells may interlock and become packed in the smaller capillaries in such a manner as to cause stasis, edema, and congestion, thereby producing added sickling because of anoxemia (1). The pathological lesions in bone have been described by Diggs, Pulliam, and King (11) as degenerative processes (thrombosis, infarction, necrosis, hemorrhage) and reparative processes (hyalinization, fibrosis, abnormal calcification and new bone formation). They are similar to those found in circulatory disturbances due to trauma or caisson disease (16, 17, 27, 28).

The purpose of this report is to describe the roentgenographic findings in 26 cases of sickle-cell anemia in adults seen at the Presbyterian Hospital in the City of New York since 1930. Particular attention is given to the appearance of the bones, although the prominent clinical features and blood findings are presented in tabular form (Table I). Two cases are reported in some detail. One (Case 6) illustrates the generalized osteosclerosis (Fig. 9) previously described (11, 19, 20). The other (Case 8) seems to be unique in that what appears to have been an infarction in the body of a vertebra was followed with serial films (Fig. 7) from its inception to a healed

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TABLE I: CLINICAL FINDINGS IN 26 CASES OF SICKLE-CELL ANEMIA*

Case No., Sex, and Age	R. B. C. (million)	Hb. (gm., 100%)	Sickling, %	Leg Ulcer	Abd. Crises	Valvular Dis.	Operation	Autopsy	Additional Diagnoses	Chest	Skull	Spine			Long Bones			Remarks
												Cervical	Thoracic	Lumbar	Pelvis	Femur	Tibia	
1. M. 19	3.0	9.5	80	+	+	..	0	...	Bronchopneumonia	X-1	X	X	X	X	Generalized demineralization
2. M. 20	2.5	8.4	95	0	0	..	0	...	0	X-1	X	X	X	..	X	X	X	Generalized demineralization
3. M. 20	3.7	6.2	100	0	0	..	0	...	Lobar pneumonia	X	Generalized demineralization
4. M. 25	3.2	13	20	+	0	..	0	...	Pulmonary tbc., syphilis	X	Generalized demineralization
5. M. 25	2.9	9.5	60	0	0	..	Bone graft—hip	...	0	X-1	..	X	X-2	X-2	X-3, 4	X	X	Generalized demineralization
6. M. 26	3.3	10	100	+	+	..	Liver biopsy	...	0	X	..	X-2	X-2	X-2	..	X-3	X-3	Asplenic necrosis of rt. hip
7. M. 29	4.8	9.9	20	0	0	..	0	...	Pleural effusion; syphilis	X-1	Infarct, tarsal navicular. Osteosclerosis; dense spleen
8. M. 32	3.4	10	80	0	+	..	Exploratory laparotomy	...	0	X	..	X-2	X-2, 4	X-2, 4	X	X	X	Generalized demineralization
9. M. 32	2.6	8.5	100	+	0	..	Splenectomy	...	Rheumatic heart disease; syphilis	X-1	X	X	Generalized demineralization
10. M. 34	3.6	10	90	0	+	0	Exploratory laparotomy	Yes	0	X-1	Generalized demineralization
11. M. 42	4.0	14	60	0	+	..	Exploratory laparotomy	...	0	X	X	Generalized demineralization
12. M. 49	3.8	11	..	0	0	0	0	Yes	Pulmonary tbc.; pyelitis	X	X	Generalized demineralization
13. F. 16	3.0	9.4	98	+	+	..	0	...	0	X-1	X	..	X-2	X-2	..	X	X	Generalized demineralization
14. F. 20	2.5	6.7	75	0	0	..	Herniorrhaphy	...	Icthyosis	X-1	X	X	X	X-3	Generalized demineralization
15. F. 26	1.9	7.9	80	+	0	..	0	...	Acute hepatitis; lung abscess	X	X	Generalized demineralization
16. F. 28	4.0	12	100	0	0	..	Splenectomy	...	0	..	X	X-3	Generalized demineralization
17. F. 30	2.4	8.6	80	+	+	0	Dilatation and curettage	Yes	0	X	X-3	X-3	X	Generalized demineralization
18. F. 32	3.6	7.7	75	0	0	..	Hysterectomy	...	Fibroid uterus; syphilis	..	X	X	Generalized demineralization
19. F. 33	4.2	8.7	50	0	0	0	Hysterectomy	Yes	Fibroid uterus	X	X	X	Generalized demineralization
20. F. 38	2.75	5	100	+	+	..	Exploratory laparotomy	...	Thrombophlebitis (femoral)	X-1	X-4	X	..	X-3	X-3	Generalized demineralization
21. F. 38	2.8	10	100	0	+	..	0	...	Rheumatic heart disease	X-1	X	Generalized demineralization
22. F. 40	2.4	7.5	..	0	0	0	0	Yes	Cirrhosis; chronic nephritis	X	..	X	X	X	Generalized demineralization
23. F. 43	3.2	8	40	0	+	..	0	...	0	X-1	X	Ankylosis of hip (cause?)
24. F. 44	3.3	10	100	0	0	..	0	...	Pleural effusion	X-1	X	Generalized demineralization
25. F. 47	2.4	5.5	100	0	0	..	Cervical polypectomy	...	Lymphophathic nereum; syphilis	X-1	X	..	X-4	X	Generalized demineralization
26. F. 62	2.9	8.7	100	0	0	..	Splenectomy	...	Thrombocytopenic purpura	X-1	X	X	Generalized demineralization

*1. Cardiac enlargement. 2. Biomechanical vertebrae. 3. Patchy cortical thickening. 4. Bone infarct.

stage. A postmortem examination was made on 5 of the 26 cases.

CLINICAL MANIFESTATIONS OF THE DISEASE

Of the 26 patients, 14 were women and 12 were men. Twenty-five were Negroes. One was a native-born white man of Greek parentage. The youngest was sixteen and the oldest sixty-two. The average age for the group was thirty-one years.

The average red blood count was 3,170,000. On the basis of 16 gm. representing 100 per cent, the average hemoglobin content was 9.7 gm. or 60 per cent. The degree of sickling was determined after incubating a sealed moist preparation. Sickling was sometimes shown on direct smear, but in the majority of instances was seen only in the chamber. The percentage ranged from 20 to 100 per cent. The average was 72 per cent.

Eight of the 26 patients (approximately one-third) had either an indolent ulcer or a scar from a former ulcer on the lower third of one or both legs.

Ten patients had one or more abdominal crises, *i.e.*, attacks of severe abdominal pain accompanied by tenderness, rigidity, and signs of shock. The average white blood count was 19,000 per cu. mm. This suggested a perforated viscus so strongly in 5 of the cases that an exploratory laparotomy was done. No gross pathology was found.

In several cases cardiac murmurs were heard at various times. However, valvular disease was found in none of the 5 cases autopsied. Four of these had had apical systolic murmurs and accentuated second pulmonic sounds. It is presumed that the murmurs were hemic in origin, associated with the severe anemia.

No significant preponderance of any one associated disease was noted. Splenectomy, done in 3 cases, did not appear to influence the course of the disease.

Autopsy was performed on 5 patients. The heart was moderately enlarged in 3, with hypertrophy and dilatation of both ventricles. The endocardium was normal in all instances. Two showed pulmonary

TABLE II: POSITIVE X-RAY FINDINGS PRESENT IN 22 OF THE 26 CASES

	Cases
Enlarged heart.....	13
Generalized osteoporosis.....	11
Generalized osteosclerosis.....	1
Biconcave deformity of vertebral bodies.....	4
Patchy cortical thickening of long bones.....	7
Infarcts of bone	
Skull.....	1
Tarsal navicular.....	1
Head of femur.....	1
Head of femur and shafts of femora.....	1
Lumbar spine and shafts of femora.....	1

edema. In a third, the smaller pulmonary vessels were filled with fresh thrombi. The spleen showed profound atrophy and fibrosis in 2 cases, weighing 30 gm. and 8 gm.; masses of sickle-cells were all that remained of the cellular pulp; the malpighian corpuscles were largely missing. The pulp was strikingly hemorrhagic in the others, with pools of blood surrounding the malpighian corpuscles and great numbers of sickle-cells visible. Enlargement and marked congestion of the liver with areas of focal necrosis were noted in 2 cases. In a third, the liver was small and showed periportal fibrosis and focal necrosis. The other 2 disclosed no abnormalities. Marked hyperplasia of the bone marrow was noted in one instance. In another, in sections taken from rib and sternum, small localized areas of eroded necrotic bone trabeculae were evident, in part enclosed by fibrous tissue containing osteoclast cells.

Two patients examined at necropsy died with clinical signs of abdominal crisis. They showed marked atrophy of the spleen, but no other similarities. In the first there was great congestion of the liver sinusoids and focal necroses; also thrombi in the small pulmonary vessels. In the second, the liver was not unusual, but there were petechial hemorrhages throughout the stomach, small intestine, mesentery, pleura, and pericardium. Pulmonary edema was present.

X-RAY FINDINGS

The most frequent abnormal roentgenographic finding was cardiac enlargement. This was present in half the cases. A generalized demineralization was found in

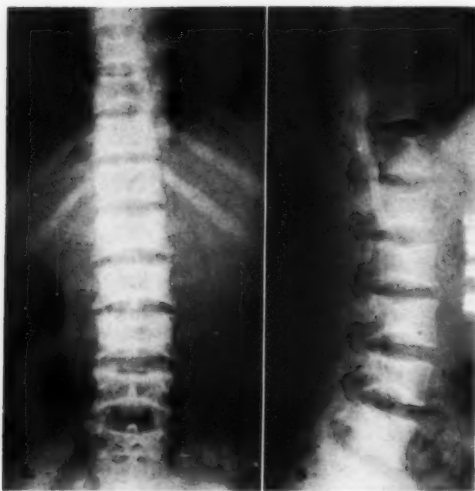


Fig. 1. The bodies of the vertebrae in sickle-cell anemia not infrequently are shortened in vertical height, widened, and show cup-shaped impressions in both the superior and inferior surfaces. This is a non-specific finding which is due to pressure by the nucleus pulposus upon weakened vertebral bodies.

almost half the cases. About 25 per cent showed definite bone infarction. The positive findings are recorded in Table II.

Heart: Thirteen of 24 patients showed cardiac enlargement. There was no characteristic configuration. Various murmurs were heard and several had been clinically diagnosed as rheumatic heart disease, but in the 5 cases which came to autopsy, as previously stated, no valvular lesion was found. This observation is comparable with a series reported by Klinefelter (18), in which 75 per cent showed an enlarged heart without evidence of valvular disease at necropsy. The enlargement is believed to be a compensatory hypertrophy and dilatation in response to the long-standing severe anemia and anoxemia.

Skull: The skull in one case showed several small areas of rarefaction which might be small areas of infarction. Except for reflecting the generalized osseous demineralization, films in 9 other cases disclosed no abnormality. The diploic widening, the thinning of the outer table, and the perpendicular striations of new bone usually found in children were not present in these adults.

Spine: Four of 16 patients with films of the spine showed an exaggerated biconcave appearance of the vertebral bodies, especially marked in the lumbar and lower thoracic region (Fig. 1). The bodies were relatively shortened in vertical height, widened, and showed marked cup-shaped impressions in both the superior and inferior surfaces. Isolated cases with iden-



Fig. 2. Localized bone infarction in the distal extremity of the shaft of the right femur (A) and of the left femur (B) in a twenty-five-year-old man with sickle-cell anemia. Patchy cortical thickening and translucent areas surrounded by a zone of increased density occur late in bone infarction. Compare with Figs. 4 and 5.

tical findings have been reported by Diggs, Pulliam, and King (11), Kraft and Bertel (19), and Leivy and Schnable (20). The deformity in these weight-bearing bones is presumably caused by the pressure of the nucleus pulposus on vertebral bodies weakened by osteoporosis. The appearance is not specific for sickle-cell anemia. It is seen in various other diseases, such as hyperparathyroidism, osteomalacia due to a diet deficient in calcium and vitamin D, in deficient absorption, as in steatorrhea, and in postmenopausal osteoporosis.

Long Bones: Patchy cortical thickening in the femur, tibia, or fibula was found in 7 of the 15 cases. Translucent areas 2 cm. in diameter, ringed by sclerotic bone,* at the distal ends of both femora were present in two cases (Fig. 2). Identical findings were demonstrable in the head of the femur, and in one case in the tarsal navicular (Fig. 3). A fifth case showed similar changes in the skull. These findings we believe represent bone infarcts. They reflect the well known thrombotic tendency in this disease as a result of circulatory stasis in the small vessels. The appearance is due to a central necrosis, surrounded by a zone of fibrosis which may be partially ossified or calcified. In one of our autopsied cases, sections of rib and sternum had this appearance. Diggs, Pulliam, and King demonstrated these findings in several of their sections of long bones.

The pathologic changes of aseptic necrosis and repair appear to be quite like those Phemister (27, 28) has observed in cases of interruption of circulation in bone following trauma and in caisson disease. Necrosis of bone results from the vascular blockage. A reparative reaction is then set up in the living bone about the dead area. Phemister has described first a fibrous tissue invasion from the surrounding living bone, which breaks down and replaces the dead marrow. The fibrous tissue in turn is transformed into new bone, which further replaces the dead bone. Small necrotic areas are usually completely replaced by new bone, but with large necrotic areas the reparative reaction may stop short of completion. There would then be a central necrotic area which in time might show patchy calcification. Surrounding this would be calcified new bone. Several reports on infarcts of the long bones due to a variety of causes, including sickle-cell anemia, have been published (16, 17, 22, 20, 33). The x-ray appearance in all is much the same (Figs. 4 and 5). When the infarct occurs in the bones about a weight-bearing joint, aseptic necrosis may be followed by a degenerative arthritis. One



Fig. 3. Bone infarction in the tarsal navicular in a man aged twenty-six with sickle-cell anemia. There is a central area of diminished density surrounded by a zone of increased density.

of our cases demonstrated this in the hip joint (Fig. 6).

Very few serial roentgenographic studies of the progressive changes occurring in a bone following the occlusion of so-called end arteries (infarction) have been made. The reasons for this are given by Phemister (27): "(1) absence or mildness of immediate symptoms resulting from simple interruption of circulation in bone, (2) roentgen signs of this condition are late in appearing and were long confused with other chronic lesions, (3) postmortem examination of the skeleton is less common and less extensive than are those of the viscera, and (4) operation affording a chance for pathologic examination is seldom indicated with most forms of aseptic bone necrosis."

The following case is reported because it presented the unique opportunity of making a serial roentgenographic study of an infarction of the body of a vertebra from its inception to a healing stage over a period of eighteen months:

A 30-year-old Negro (Case 8 in Table I) was admitted to Presbyterian Hospital Dec. 12, 1945, because of severe girdle-type upper abdominal pain which began suddenly seven hours previously. He had no nausea or vomiting. There was a history of occasional attacks of what he called rheumatism for



Fig. 4. Bone infarction of the humerus in a man aged fifty-five who had caisson disease. Views taken in external (A) and internal (B) rotation. Aseptic necrosis and reparative processes occur as in sickle-cell anemia.

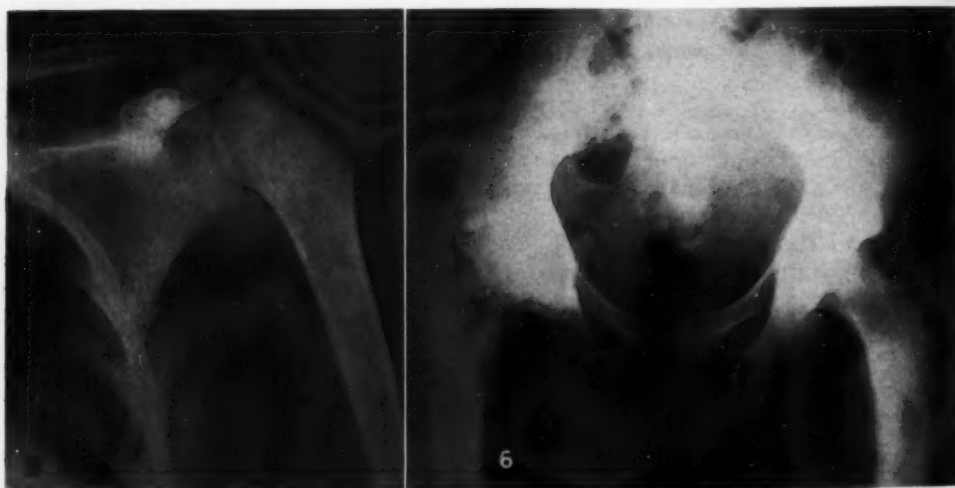
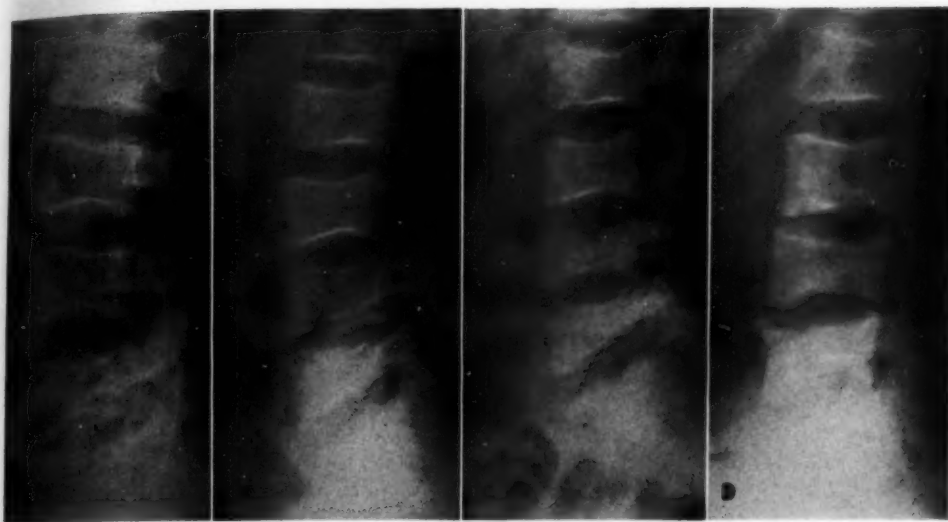


Fig. 5. Bone infarction of the humerus in a woman aged sixty-three with long-standing polycythemia vera. The lesion is not so sharply delimited as that in Fig. 4, but the patchy increase in density and irregular ill-defined translucent areas are similar.

Fig. 6. When bone infarction occurs adjacent to a weight-bearing joint, collapse of the affected bone may occur. This patient was a twenty-five-year-old man with sickle-cell anemia. Degenerative arthritis is a sequel to the aseptic necrosis. An extra-articular fusion by bone graft for a partially collapsed femoral head was done at another hospital (date not known).



many years. His abdomen was board-like and diffusely tender. No tenderness was noted over the spine. Both deep and superficial reflexes were equal and active. Temperature was 100.2°; pulse rate 90; white blood count 17,900, with polymorphonuclears 80 per cent; red cell count 3,400,000; hemoglobin 10 gm.; Kline reaction negative; erythrocyte sedimentation rate normal.

A diagnosis of ruptured peptic ulcer was made and a laparotomy was performed. No lesion was found. A wet incubated blood specimen was then examined and it showed marked sickling at twenty-four hours. The erythrocyte sedimentation rate, under oxygen, was 40 mm. at the end of an hour. The abdominal pain subsided after operation, but a swinging type of fever curve continued.

The day following operation the patient began to complain of pain in the left lumbosacral region, radiating down to the right hip. A neurologic examination disclosed marked tenderness over the spine from L-4 to S-1. Pressure on this region reproduced the pain along the posterior thighs. Paravertebral muscle spasm was present. Thigh and leg muscles were somewhat flaccid. Deep reflexes were hyperactive. Sensory examination was normal. The upper extremities and cranial nerves were normal. The clinical impression was radiculitis secondary to a lumbosacral arthropathy. Lumbar puncture disclosed normal pressure and dynamics. The spinal fluid protein, sugar, chloride, Wassermann reaction, and colloidal gold curve were normal.

Roentgenograms of the spine, pelvis, and both hips were obtained two weeks after the onset, and revealed no evidence of localized bone or joint disease (Fig. 7A). The patient's temperature and white blood count returned to normal. He con-



Fig. 7. Infarction of the bodies of L-4 and L-5 in a thirty-year-old man with sickle-cell anemia. A: Two weeks following an abdominal crisis the film shows nothing unusual. B: Eight weeks later, areas of rarefaction are present in the body of L-4 with compression of the adjacent margins of L-4 and L-5. C: Ten weeks after admission the bodies of L-4 and L-5 are still further compressed. There is an area of rarefaction in the anterosuperior portion of the body of L-5. D, E, and F: Eight months, ten months, and twelve months, respectively, after the abdominal crisis, there is almost restitution to normal in the shape of the affected bodies. A homogeneous increase in density of both L-4 and L-5 is present in the last film and there is narrowing of the intervertebral disk space between L-3 and L-4 and between L-4 and L-5.

The occurrence of bone infarction in this case at the time of the abdominal crisis might or might not be fortuitous.



Fig. 8. Bone infarction in the distal extremity of the shaft of the femur, in the same patient who had infarcts of the lumbar vertebrae, shown in Fig. 7. An identical appearance was present in the opposite femur.

tinued to complain of low back pain, particularly over L-4, radiating to both hips. Roentgen examination of the lower lumbar spine was repeated Feb. 8 and Feb. 20, 1946 (eight and ten weeks after the onset of the illness), revealing for the first time x-ray evidence of a disease process in the bodies of L-4 and L-5 (Fig. 7, B and C). Areas of translucency were present in the body of L-4 and in the upper anterior margin of L-5, adjacent to the intervertebral disk. The adjacent margins of L-4 and L-5 were compressed. The disk spaces were not narrowed. These findings were interpreted as being due to infarction, although other possibilities such as tuberculosis, osteomyelitis, myeloma, and lymphoblastoma were entertained. Roentgenograms of both knees including the lower femora were taken. They showed patchy cortical thickening and localized translucent areas surrounded by dense bone, which were consistent with old infarcts (Fig. 8, A and B).

Blood chemistry findings were: serum calcium, normal; serum alkaline phosphatase, 5.6 Bodansky units; acid phosphatase, 3.2 Gutman units; serum protein and A/G ratio normal. Uric acid was normal. Bence-Jones protein was not found in the urine. Blood culture gave no growth. A chest film revealed no sign of a tuberculous process. A sternal marrow biopsy showed no evidence of leukemia or myeloma. No primary neoplasm was found in the gastro-intestinal or genito-urinary tract. A hemolytic streptococcus agglutination test was negative.

Treatment consisted of diathermy and passive exercises. At no time were sulfonamides or antibiotics administered. About three months after the onset of the illness, the patient was asymptom-

atic. The sedimentation rate (under oxygen) was normal. Films of the spine on March 15, 1946, showed no change in the appearance of L-4 and L-5 on comparison with films of Feb. 20. A Taylor spinal brace was applied as a precaution against further compression of the vertebral bodies. The patient returned to work eight months after the onset of the illness, free from pain. The brace was discarded, at the patient's request, after his return to work. Re-examinations at eight months (Fig. 7D), ten months (Fig. 7E) and one year later (Fig. 7F) showed an increase in density of the bodies of both L-4 and L-5, with disappearance of the translucent areas adjacent to the disks. Compression of the bodies was no longer present. A slight narrowing of the disk spaces was found. Some lipping or spur formation can be seen at their anterior margins. The anteroposterior views are not reproduced because the bone changes cannot be seen distinctly except when viewed stereoscopically. A follow-up examination twenty-two months later showed no appreciable change in the lumbar spine. The patient has continued to work and has been free from symptoms.

The roentgenographic appearance of the bones in some cases (11, 19, 20) of sickle-cell anemia in adults suggests a marked degree of generalized bone sclerosis with narrowing of the medullary cavities and apparent endosteal bone production. An explanation for this generalized, advanced degree of bone sclerosis, occurring relatively infrequently, as contrasted with the usual occurrence of a generalized demineralization and widened medullary cavities is not obvious. It appears to have no direct relation to the severity or duration of the anemia. It occurs infrequently and is not the usual manifestation of this disease. Only one case in this group of 26 adult sickle-cell anemias showed a pronounced degree of bone sclerosis:

The patient was a 26-year-old Negro (Case 6 in Table I) with polyarthritis, abdominal pains, ulcers of the lower legs, and exertional dyspnea intermittently for twenty years. Physical examination disclosed a mitral systolic murmur, healed ulcers of both ankles and an ulcer of the right foot, pes cavus on the left, which the patient attributed to his having to walk on the ball of his foot for many years because of an ulcer of the heel. Laboratory findings were as follows: red blood count 3,300,000; hemoglobin 10 gm., white blood count 7,500. Wet incubated blood specimen showed 100 per cent sickling at twenty-four hours. The urine urobilinogen was 2 plus. A liver biopsy showed congestion of the simu-

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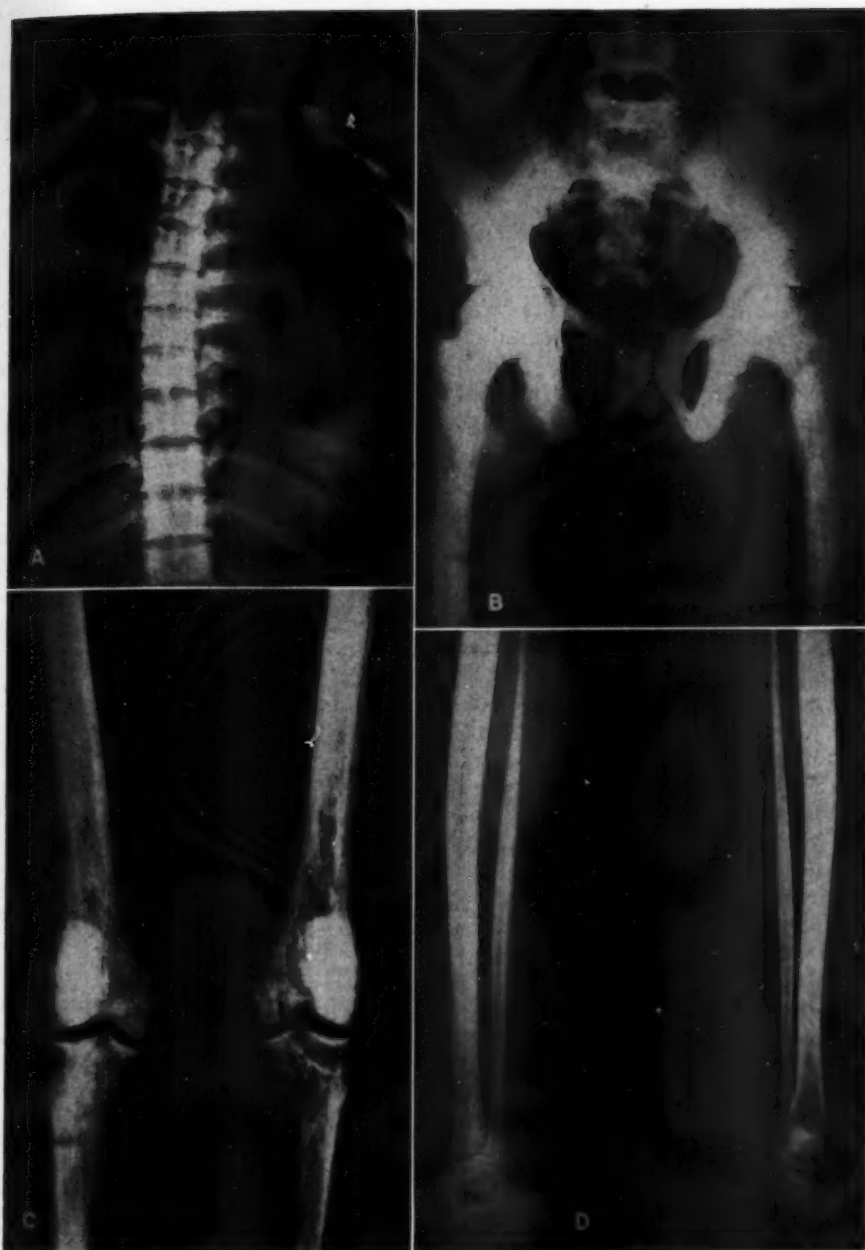


Fig. 9. Generalized osteosclerosis with narrowing of the medullary cavities in a man aged twenty-six with sickle-cell anemia. This is an unusual manifestation of the disease, occurring only once in this series of 26 cases, whereas generalized osseous demineralization was present in almost half the group. Equally unusual is an abnormally dense spleen (A), presumably containing a large amount of iron and calcium. The majority of patients with sickle-cell anemia are found to have a very small spleen at necropsy. There is no apparent correlation between size and mineral deposit. Infarct of the tarsal navicular in this patient is shown in Fig. 3.

soids and evidence of considerable hematopoiesis. Roentgenograms (Fig. 9, A-D) show a diffuse osteosclerosis, biconcave deformity of the vertebral bodies, patchy cortical thickening of the long bones, and infarcts in the tarsal navicular and calcaneus. An additional finding is a marked increase in density of the spleen, presumably because of a high iron and calcium content (Fig. 9A).

SUMMARY

The x-ray findings in 26 adults with sickle-cell anemia reflect the long-standing anemia and the peculiar thrombotic tendency in this disease. The most frequent abnormal finding was cardiac enlargement. The abnormal bone findings in this group consisted of the following: (1) generalized osseous demineralization, 11 cases, (2) a biconcave deformity of the vertebral bodies which is believed to be the result of bone softening, 4 cases, (3) patchy cortical thickening of the long bones, 7 cases, which might be due to infarction, (4) localized bone infarcts, 5 cases, not unlike those seen in caisson disease.

Four of the 26 cases showed nothing abnormal on the films. Radial striations and unusual diploic widening were not present in the skull. One case (Case 20 in Table I) did show a patchy cortical thickening consistent with infarctions.

The clinical features and laboratory findings of the 26 cases reviewed are summarized in Table I.

A unique case is reported in which a serial roentgenographic study was made of an infarction of two vertebral bodies from its inception to a healing stage.

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SUMARIO

Drepanocitemia en Adultos: Hallazgos Radiográficos

Los hallazgos radiográficos en 26 adultos con anemia por células falciformes reflejan la prolongada anemia y la peculiar tendencia trombótica de la enfermedad. El hallazgo anormal más frecuente consistió en hipertrofia cardíaca. Las anomalías óseas descubiertas en el grupo comprendieron: (1) desmineralización generalizada, 11 casos; (2) deformidad bicóncava de los cuerpos vertebrales, aparentemente debida a reblandecimiento óseo, 4 casos; (3) placas de espesamiento cortical de los huesos largos, posiblemente debido a infarto, 7

casos; (4) infartos óseos localizados algo parecidos a los observados en la enfermedad de los buzos (*caisson*).

En 4 de los 26 casos la radiografía no reveló nada anormal. En el cráneo no había estrías radiales ni inusitado ensanchamiento del diploe. Un caso sí mostró placas de espesamiento cortical compatible con infartos.

Comuníquese un caso único en el que se hizo un estudio radiográfico seriado de un infarto de dos cuerpos vertebrales desde la iniciación hasta la etapa de curación.



Polyostotic Fibrous Dysplasia (Albright's Syndrome) and Its Comparison with Dyschondroplasia (Ollier's Disease)

A Correlation of the Radiological and Pathological Findings¹

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THE PURPOSE OF this paper is to correlate the roentgenologic, pathologic, and clinical findings in polyostotic fibrous dysplasia, as indicated in a report of four cases; to present the essential roentgenologic and pathologic variations from dyschondroplasia (Ollier's disease); and to point out certain roentgenologic variations between these two diseases and other conditions of fibrocystic appearance with which they may sometimes be confused.

HISTORICAL NOTE

As early as 1922, Weil (1) reported a case in a nine-year-old girl with sexual precocity, pigmentation, and marked skeletal changes, as an "uncommon type of osteopsathyrosis." A fairly large number of such cases were subsequently reported (2-9), but these were all isolated examples appearing under many different headings. Almost simultaneously, McCune (10) described a case, and Albright, Butler, Hampton, and Smith (11) presented five instances in which this syndrome was recognized as a definite entity. It was left to Lichtenstein (12), in 1938, to separate the bone lesions occurring in this condition from other diseases with which it was confused and to provide the name polyostotic fibrous dysplasia. Up to that time the disease had been considered a particular variant of generalized osteitis fibrosa cystica (von Recklinghausen's disease), and many useless operations were performed in search of enlarged parathyroid glands.

Differentiation of polyostotic fibrous dysplasia from other similar appearing conditions was worked out by many authors: Lichtenstein and Jaffe (13), Albright and associates (11, 14), Dockerty and others (15), who stressed the presence of skeletal and, less often, extraskkeletal changes, developing in childhood. In some instances these changes progressed gradually from a single bone to many, often showing a tendency to a unilateral distribution. In others, bilateral lesions occurred with widespread involvement of bones of extremities, pelvis, and skull. Patients with such extensive disease often show abnormal patches of pigmentation of the skin and, in the case of females, sexual precocity (Albright's syndrome). Incomplete forms were also seen, in which the patient failed to exhibit one or other of the extraskkeletal features. We shall not enter into the discussion here of whether or not the term "incomplete form of fibrous dysplasia" should be applied to such a solitary localized lesion in an otherwise healthy individual.

CLINICAL MANIFESTATIONS

Clinically, fibrous dysplasia of bone is insidious in its onset. The development of osseous variations is usually slow, and there may be little, if any, associated pain or disability; later on, pain may become a prominent symptom. The process may, however, be well developed throughout the bony structures of the body, and yet go

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unsuspected; not until there is interference with proper function or pathological fractures occur, may the condition be suspected. The diagnosis is usually made by roentgen examination.

The process is slow but unrelenting; the lesions may be large and fully developed in one bone and in their initial stage in another. Weakening of the bone may lead to spontaneous fractures, but the gross deformities from bending of the bones seen in generalized osteitis fibrosa cystica (von Recklinghausen's disease) do not develop.

The earlier lesions may give a clearer insight into the true nature of the disease. This may be recognized by the distinctive findings produced by the progression of the pathological process as it involves normal bone.

ROENTGENOLOGIC FINDINGS

The earliest manifestation of polyostotic fibrous dysplasia is the formation of tiny "cyst"-like areas within the cortical structure, often no more than a few millimeters in diameter when first observed (Fig. 7). These rounded areas are not true cysts but are fibrous tissue deposits which cause replacement of the normal trabecular structure by a homogeneous granular appearance devoid of bony architecture. As they increase in size, they cause pressure on the surrounding cortical bone; if the area of involvement is nearer the medullary side, pressure of the growth, progressing in the direction of least resistance, will be inward toward the medullary canal, giving rise to a curved crescentic inner border; if least resistance is offered by the outer cortical layer, localized expansion will be outward (Figs. 2 and 8). Extremely superficial lesions often cause rounded, "blister"-like elevations on the surface of the bone. These rounded "cyst"-like areas of unilateral expansion occurring within the cortex of the shafts of the long bones are the most diagnostic lesions of this condition, but they are not absolutely characteristic.

As these rounded areas become larger, they may encroach upon each other, giving the appearance of pseudo-expansion;

the bony outline may assume a knobby appearance (Figs. 9 and 10).

Shell-like thinning of the cortex may result, causing marked weakening of the bone.

Spontaneous fractures may follow weakening of the bony structure, but these always heal readily by new bone formation (Fig. 3).

No evidence of periosteal elevation is seen in the roentgenogram unless in response to fracture.

Osteoporosis does not occur, as in generalized osteitis fibrosa cystica; so there is no softening of the bone and bending deformities are not produced.

Varying degrees of *sclerosis of the involved bones* occur under certain conditions. The more cancellous the bony tissue at the site of involvement, the less is the resistance offered to the advancement of the process and the less the resulting sclerosis; the more dense the structure of the bone involved, the more sclerotic the resulting bony structure. In the region of the greater trochanter, where the bone is quite cancellous, there is little resistance to the progression of the process and little if any sclerosis. When, on the other hand, the disease occurs in the petrous portion of the temporal bone, sclerosis is most marked. Indeed, the rounded "cyst"-like areas seen in other bones are entirely absent here. (Fig. 13).

Widening and elongation of the shaft of the bone occur as a result of the hyperplastic process within the medullary canal and interstitial spaces (Figs. 9 and 10).

The bone lesions have a tendency to *unilateral involvement* (Figs. 1-5). Even when the cranial bones are involved, this tendency to unilateral involvement is preserved. Later on, both sides may become involved. In the polyostotic form, the disease shows varying degrees of development in the same or adjacent bones.

Bony lesions of the type described may be associated with *sexual precocity in the female and pigmented areas of the skin*, other manifestations of Albright's syndrome (Figs. 1, 6, 15, 17).

The process may seem to become quiescent, but the normal bony architecture is never restored. Spontaneous fractures occur due to the weakened condition of the bone from the multiple areas of fibrotic replacement. The bone becomes structurally weak, not soft; there is no osteoporosis. For this reason, the bones may break, but they do not bend or form gross deformities as in von Recklinghausen's disease.

In the bones of the hands and feet (Figs. 4 and 11), the process becomes confluent, filling the entire medullary canal and causing rather uniform symmetrical enlargement of the bony structures. There is no subperiosteal granulation as in tuberculous dactylitis, and all the bones are usually involved.

In the skull (Figs. 5, 13, and 18), when the calvarium is involved, there is irregular thickening of the outer table: if the lesions are superficial, "cyst"-like areas, usually large and quite spherical, may appear. The inner table presents a dense resisting barrier preventing encroachment upon the cranial cavity. When the basal bone structures are involved, the dense character of the cortical bone prevents expansion, and an appearance of dense bone sclerosis results. The picture is very similar to that of leontiasis ossea. However, we do not believe that leontiasis ossea is a localized type of fibrous dysplasia, but rather that it is an ossifying fibrosis. In the flat bones of the pelvis, there is a minimal amount of resistance, and the fibrous "cyst"-like areas, unrestrained in their growth, assume almost circular form.

ETIOLOGY

The etiology of polyostotic fibrous dysplasia is not well understood. While Lichtenstein and Jaffe (13) favor a congenital disturbance of the activity of the undifferentiated fibrous bone-forming mesenchyme, Albright (14) considers damage to the hypothalamus region as the cause, an idea which Goldhamer (5) vaguely expressed in 1934 when he spoke of an involvement of the base of the skull. Freund

(8) advanced the opinion of an anomaly of the sympathetic system, and Uehlinger suggested that a temporary hormonal disturbance, particularly of the adrenals, seemed not unlikely. The calcium-phosphorus balance is not disturbed; the serum phosphatase is sometimes increased. These are the only significant findings in the blood chemistry which may be of value in differential diagnosis.

MICROSCOPIC FINDINGS IN POLYOSTOTIC FIBROUS DYSPLASIA

(Plate I, A and B)

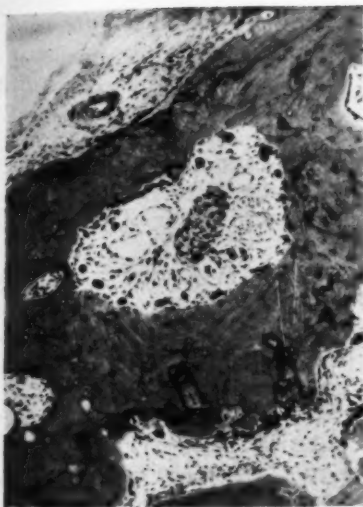
There is an *osteoclastic absorption of the bony structure followed by fibrosis of the bone marrow in the cortical and subcortical areas. The process starts in the haversian canals and spreads to the surrounding structures.* The cortex becomes trabeculated and thinned, and many trabeculae of the spongy bone are resorbed, while a more or less loose fibrous tissue fills in the spaces. This fibrous tissue may contain many capillaries surrounded by edema. Loosely arranged stellate cells may be seen next to dense fibrous, almost scar-like, tissue. We have never observed true cyst formation. In many areas the spaces between the small star-like cells are occupied by collagen, which may become very dense and undergo calcification. In only a few places are there moderate collections of osteoclasts which were left after destruction of the bone. The fibrous tissue contains many bizarre, newly formed, slender bone trabeculae, the coarsely and irregularly calcified centers of which appear coated by broad osteoid zones with entrapped threads of connective tissue. This is observed particularly in the dense fibrous areas close to the cortex, where the deposition and fusion of collagen masses so rapidly occur that their calcification becomes inadequate and extremely wide osteoid tissue surrounds irregularly calcified centers. In contrast to other authors, we observed a fairly remarkable osteoclastic resorption of the newly formed slender trabeculae. Osteoclasts were seen in calcified and osteoid tissue alike.

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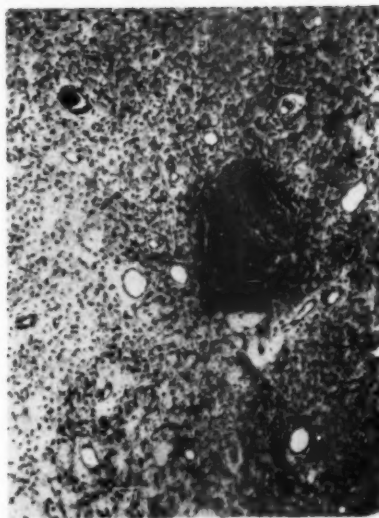
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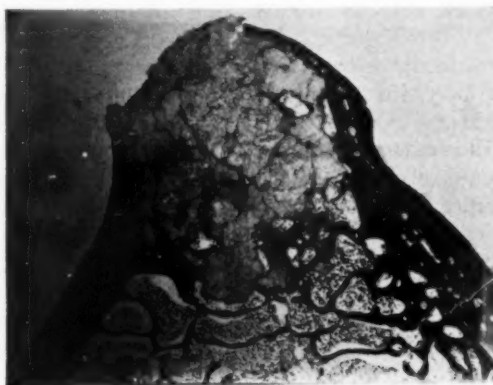
PLATE I



A. Polyostotic fibrous dysplasia. Cortex of femur covered by normal periosteum shows osteoclastically dilated haversian systems, the contents of which consist of loose fibrous tissue with expanded capillaries. Slight periosteal bone deposition.



B. Polyostotic fibrous dysplasia. Area of the interior of a lesion. Fibrous tissue rich in collagen showing many dilated thin-walled capillaries and an island of irregularly calcified fibrous bone, the latter surrounded by a dense fibrous zone.



C. Ollier's disease. Blister-like expansion of the middle shaft of the femur. Cyst-like cavity filled with hyaline cartilage. The thinned cortical plate is perforated by protruding cartilage. The cartilage borders normal mixed bone marrow.

Microscopic study of the roentgenographically normal bone next to the lesions offers the opportunity to observe the initial changes in the marrow. The process starts with a thickening of the endosteum and a concentrically progressing loose fibrosis of the bone marrow, with congested capillaries and osteoclastic resorption of the bone trabeculae. Microscopically, a moderate periosteal bone deposition was noted, even though no periosteal new bone formation could be detected in the roentgenogram. In a few instances we could also observe small islands of hyaline cartilage embedded in the characteristic tissue of polyostotic fibrous dysplasia.

DIFFERENTIATION FROM OTHER "CYST"-LIKE DISEASES OF BONE

Polyostotic fibrous dysplasia is easily differentiated from other types of bone involvement if the extraskkeletal lesions, abnormal skin pigmentation, and sexual precocity in the female are present. When fully established, the roentgenographic findings are in some respects similar to those of generalized osteitis fibrosa cystica (von Recklinghausen's disease). In the developing stage there is one outstanding difference: in generalized osteitis fibrosa cystica there is extreme osteoporosis, due to the hyperparathyroidism which is invariably present; in polyostotic fibrous dysplasia this is lacking. Osteoporosis is responsible for the extreme degree of deformity from bending of the bones which sometimes occurs in generalized osteitis fibrosa cystica; in polyostotic fibrous dysplasia, deformity, if any, is local, due to enlargement of individual lesions; the bones become weak from thinning of the cortex and may fracture, but bending as a result of softening is not encountered. In fact, it is amazing how little change in appearance in the structure of the individual is evident with extensive bony involvement. If the process has not gone too far in replacement of the normal bony structure, lack of osteoporosis would indicate that it probably is due to polyostotic fibrous dysplasia; whereas, the pres-

ence of extreme osteoporosis would suggest generalized osteitis fibrosa cystica. When there is extensive involvement, it may be impossible to judge the degree of osteoporosis without observing the other bones of the skeleton, especially the skull and the spine. There is no periosteal reaction seen roentgenographically in polyostotic fibrous dysplasia except where pathological fractures have taken place. Pathological examination will readily establish the differentiation.

In polyostotic fibrous dysplasia the expanded areas in the cortical and subcortical regions are not true cysts, but are filled with fibrous tissue; they are sharply delineated by the bony structure. The intervening bone appears normal, and there is no osteoporosis or other evidence of parathyroid involvement. In generalized osteitis fibrosa cystica, the vacuolated areas are true cysts in the bone, arising also in the cortical structures, having at times definite lining membranes of fine fibrous tissue and containing clear straw-colored or blood-tinged fluid. The remainder of the bony structure shows osteoporosis, and between the trabeculae there is extensive fibrous tissue. These changes are no doubt due to hyperparathyroidism caused by either parathyroid hyperplasia or tumor. It is obvious then that from the roentgenogram alone absolute differentiation between these two conditions may not be possible. Correlation with clinical findings and mineral metabolism, however, should establish the diagnosis. In any case in which the differentiation is not clear, a biopsy from some readily accessible portion of a bone lesion would seem advisable before undertaking operative procedure in search of enlarged parathyroid glands.

Solitary areas of fibrosis are encountered in bone presenting a microscopic picture identical with that seen in fibrous dysplasia. Such areas often show no change over long periods of time; it is questionable whether they represent solitary foci of fibrous dysplasia. Certainly, unless there are other associated clinical symptoms,

such as precocious menstruation and skin pigmentation, the diagnosis should not be made.

A number of other fibrotic and cyst-like appearing lesions in bone must be differentiated.

Ordinary *unicameral bone cyst* is solitary, occurring in individuals under twenty years of age, in the diaphyseal ends of long bones, in close relationship to the epiphyseal plate. The lesion is central in origin and is always attended by true symmetrical expansion of the shaft of the bone. It is thought to have its origin from local hemorrhage from some sort of trauma to the bone. Microscopically, such lesions are true cysts, filled with fluid and at times lined by a thin connective-tissue membrane.

Giant-cell tumor, which the lesions of polyostotic fibrous dysplasia may resemble, usually occurs in individuals over twenty years of age; it is solitary and shows expansion but is traversed by many strand-like barriers of bone, giving an appearance of trabeculation. It occurs at the end of a long bone. Microscopically, it is characterized by formation of giant cells of a particular type.

Solitary expanding lesions involving the ends of long bones in adults may be due to *non-ossifying fibroma*. Microscopically, they appear similar to other fibromas occurring in bone, except that the fibroblasts show no tendency to bone formation.

Enchondroma, a benign cartilaginous tumor, is central in location, producing true expansion like a bone cyst, but such lesions are usually multiple and occur in the small bones of the fingers and toes. If they are larger, they usually occupy a position in the shaft and may take on a multiple lobulated appearance.

Cyst-like areas of degeneration may occur in the long bones in *Paget's disease*, but these are usually central and are in the mid portion of the shaft. Typical manifestations of Paget's disease seen in other bones will aid in differentiation. The skull in Paget's disease may give an appearance resembling fibrous dysplasia of bone, but

here again lesions found in the long bones will aid in differentiation.

DYSCHONDROPLASIA (OLLIER'S DISEASE)

Differentiation from other rarer types of similar appearing bone lesions, such as Ollier's dyschondroplasia, has not been widely discussed in the literature. In this condition roentgen examination discloses irregularly distributed rarefied areas in the diaphyses showing unilateral expansion of the metaphyses and thinning of the cortex (Figs. 21 and 22). Either a single bone may be involved or there may be manifestations of involvement in a number of bones. There is pressure absorption but no periosteal reaction or other indication of new bone production.

This similarity, although superficial, leads some authors to believe that fibrous dysplasia and Ollier's disease are different stages of the same condition. The origin of Ollier's disease in the cancellous structure in the region of the metaphysis and the development of the lesions of fibrous dysplasia in the cortical regions of the shafts of the long bones should aid materially in differential diagnosis. Moreover, in Ollier's disease, the roentgenogram reveals *longitudinal bony strands of preserved bone trabeculae radiating fan-like at the ends of the involved bones, with the rarefied areas caused by cartilage deposits in between. Small heavily calcified globular bodies are observed within these mottled areas.* Both of these findings are very important in differential diagnosis. The condition arises in early childhood during the period of bone growth. As the bone develops, the inhibition of bone growth in the involved portions of the diaphyses and the continuous normal bone development in the unaffected areas result in marked deformity and shortening of the affected extremity. As bone growth continues, the cartilaginous rests may be displaced farther from the metaphyseal region into the shaft.

Microscopic Findings in Dyschondroplasia (Ollier's Disease) (Plate I, C): Pathologic examination shows the rarefied areas to be

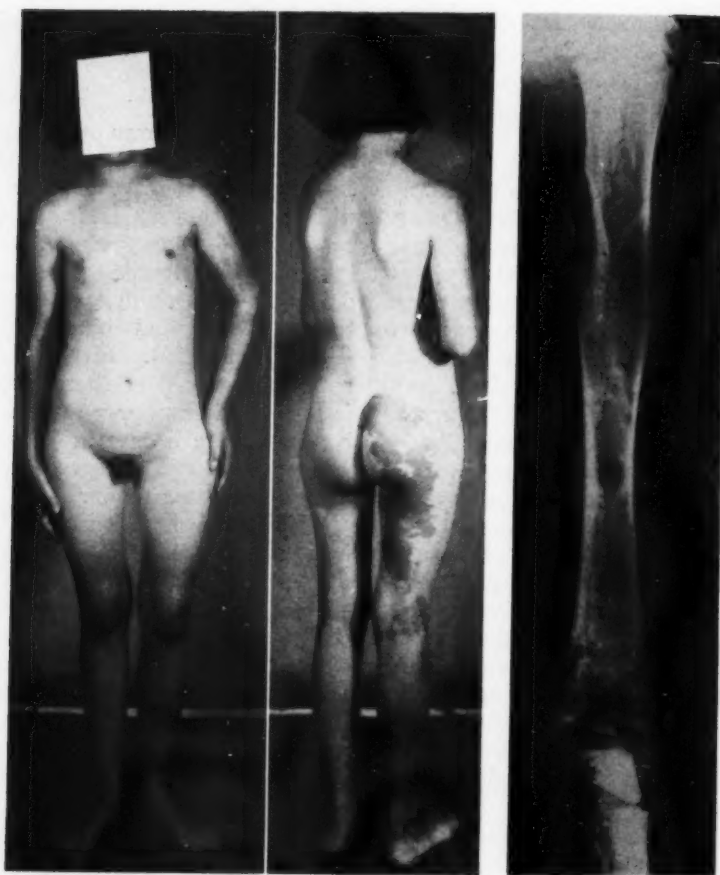


Fig. 1. Case 1: Large pigmented areas over right side, back, and breast, with bone lesions and precocious menstruation, constituting Albright's syndrome.

Fig. 2. Case 1: Intracortical "cyst"-like lesions in the humerus extending crescentically in toward the medullary canal. This is the most characteristic lesion of polyostotic fibrous dysplasia of bone.

due to cartilaginous proliferation remaining within the bony structure, causing faulty bone growth, not to fibrous tissue deposits or to fluid-containing cysts. All of the roentgenologic findings at once become clear.

Cortical bone is expanded and thinned and may even be completely destroyed by a protruding mass of immature, irregularly arranged cartilage. The periosteum which covers some areas of cartilage appears normal, with no tendency toward bone deposition. The whole area of cartilage is surrounded by normal bone so that the cartilage in some places meets unchanged

fat bone marrow and mixed bone marrow. There is not the slightest tendency to the development of fibrous marrow. The surrounding bone includes islands of cartilage. There is a definite polymorphism of the cartilage cells, many of which show a vacuolated cytoplasm with eccentrically displaced nuclei. Other cartilage cells appear extremely small, with pyknotic nuclei. The cytoplasm is sometimes granular. The intercellular substance is occasionally abundant. There are dystrophically calcified parts of cartilage which are undoubtedly the roundish calcified bodies seen in the roentgenogram. Thin plates



Fig. 4

Fig. 5

of bone are observed in the mass of cartilage.

CASE REPORTS

CASE 1 (Figs. 1-5): F. M. T., white female, age 12, was admitted to St. Mary's Hospital, March 13, 1943, complaining of weakness and limping of the left leg. The condition dated from the age of eight years. A diagnosis of poliomyelitis was made elsewhere in September 1942 and reaffirmed in February 1943. No previous roentgen examinations were made.

Roentgenologic examination revealed a large well developed cyst-like area involving the upper end of the shaft of the femur near the greater trochanter. The bone was expanded in a manner somewhat similar to that produced by a unicameral cyst. Owing to the location in the region of the greater trochanter there was some deformity. There was no periosteal reaction. Later a pathological fracture occurred at this site, which united promptly with new bone formation, without affecting the character of the "cyst"-like area. Owing to the nature of the lesion, x-ray studies of other bones were undertaken, and the diagnosis of polyostotic fibrous dysplasia was made. In the left foot there was involvement of all of the metatarsal and phalangeal bones with broadening of the entire bony structure. Clinically the foot revealed relatively slight deformity in relationship to the roentgen appearance of bony enlargement. There was no evidence of other involvement in the lower extremities at this time. Later, small initial lesions appeared in the left tibia and fibula.

Examination of the upper extremities showed ex-



Fig. 3. Case 1: Fibrocystic lesions in left femur, the initial site of bony involvement. Pathological fracture healed readily.

tensive involvement of the entire shaft of the left humerus, with some tendency to irregular expansion; there was no evidence of osteoporosis or periosteal reaction. In the lower end of the left radius were small rounded cyst-like areas representing early lesions *within* the cortex. Strangely enough, the metacarpals and phalanges of the left hand showed



Fig. 4. Case 1: Left hand showing extensive involvement of the bony structures; the entire shafts enlarged. Right hand uninvolved.

Fig. 5. Case 1: Involvement of left petrous bone. Only the bony structures on the left side of the body were involved in this case, even in the skull.



Fig. 6. Case 2: Large pigmented areas covering the greater portion of the back, side, and chest of an adult male, with generalized polyostotic fibrous dysplasia (Albright's syndrome).



Fig. 7. Case 2: Tiny lesion starting in cortex of radius, evidence of "earliest" involvement. Almost all of the other bones showed extensive involvement by polyostotic fibrous dysplasia.

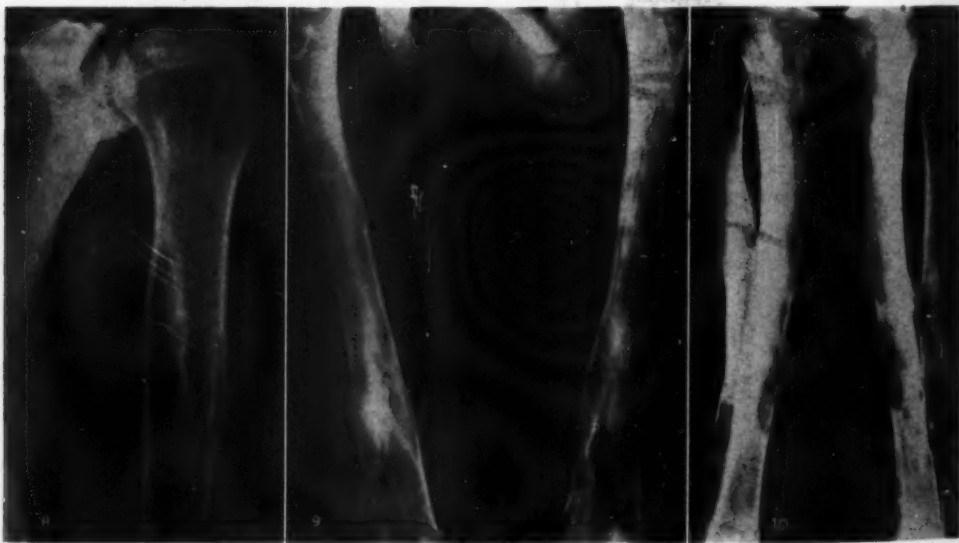


Fig. 8. Case 2: Crescentic lesions in shaft of humerus, most characteristic of polyostotic fibrous dysplasia.

Fig. 9. Case 2: Extensive involvement of both femurs, causing thick, knobby appearance, with thinning of the shafts but no bending deformity.

Fig. 10. Case 2: Bones of both legs similarly involved; somewhat elongated because of hyperplastic process.

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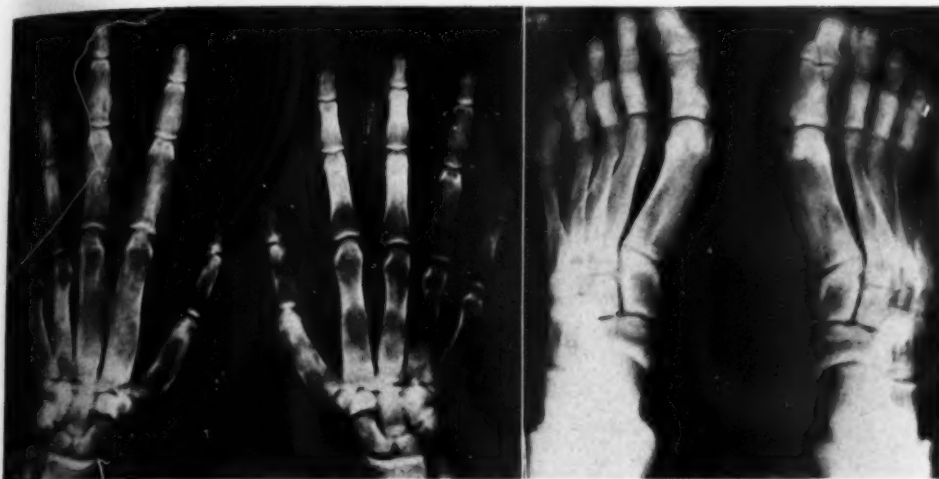


Fig. 11. Case 2: Hands and feet showing extensive involvement of all bones.

the disease well established before the left forearm was involved.

The first manifestation of skull involvement occurred in the petrous bone on the left side. It never assumed a "cyst"-like appearance.

Menstruation had begun at nine years of age and pubic and axillary hair was well developed. Large areas of skin pigmentation were present on the right buttock, back of the right thigh, and above the right breast, thus completing Albright's syndrome. It is interesting that the pigmented areas were confined to the side opposite the bony involvement.

There was some elongation of the left lower extremity, owing to the hyperplastic nature of the disease, which probably accounted for the limp.

The patient has been under observation at intervals up to the present time, showing gradual progression of the disease. Radiation therapy was equally ineffectual in the early and the well developed lesions.

Hematological Examinations: 3-15-43: 8,650 leukocytes; 4,300,000 erythrocytes; 13.0 gm. hemoglobin.

Blood Chemistry: 3-16-43: Non-protein nitrogen 34 mg. per 100 c.c.; blood sugar 80 mg. per 100 c.c.; total inorganic phosphorus 7.2 mg.; serum inorganic phosphorus 2.9 mg.; phosphatase 4.3 Bodansky units (normal for children, 5-12 units).

3-23-43: Total inorganic phosphorus 9.2 mg.; serum inorganic phosphorus 3.8 mg.; phosphatase 5.4 Bodansky units; calcium 7.7 mg.

3-24-43: Cholesterol 200 mg.; Bence-Jones protein, negative; total inorganic phosphorus 7.6 mg.; serum inorganic phosphorus 3.0 mg.; phosphatase 4.6 Bodansky units; tourniquet test, normal; clotting time 3 1/2 min.

CASE 2 (Figs. 6-13): C. U., white male, age 25

years, had no previous knowledge of his condition and served in the U. S. Army Air Force for three years during the war, completing many missions over enemy territory. He sought medical aid a number of times for skin pigmentation and for a dull pain in his right leg, but the condition was not recognized and treatment was ineffective. Not until April 1946, after discharge from the army, when roentgen examination was made for injury to the ankle, was the true nature of the disease discovered.

The patient was then admitted to St. Mary's Hospital for complete physical and roentgen examination. Large areas of pigmentation covered the greater portion of his back and chest. Smaller areas were present on the inner aspect of the left arm and elsewhere.

Roentgen examination of the bony structures revealed extensive bilateral involvement of almost all of the long bones of both upper and lower extremities, scapulae, pelvis, and skull. The long bones were so extensively involved that the "cyst"-like areas impinged upon each other, giving a knobby appearance to the bone. The cortex in places was paper-thin, but there was no osteoporosis and the bones retained their normal contour; there was no evidence of weakening or bending of the bones. The metatarsal and metacarpal bones and phalanges of all extremities had the broadened appearance, with absence of trabeculae, usually seen in this condition. The flat scapulae and iliac bones showed rounded circinate areas of decreased density with well defined margins of bone sclerosis.

The bony structures of the skull appeared thickened and sclerotic, especially in the basal region, with "cyst"-like areas of destruction within thickened Paget-like bone.



Fig. 12. Case 2: Pelvis showing ring-like structures surrounding areas of sclerotic appearing bone, characteristic of the disease.



Fig. 13. Case 2: Extensive involvement of skull from polyostotic fibrous dysplasia, showing dense sclerosis of compact bony structures at base and tendency to cystic formation over the calvarium.

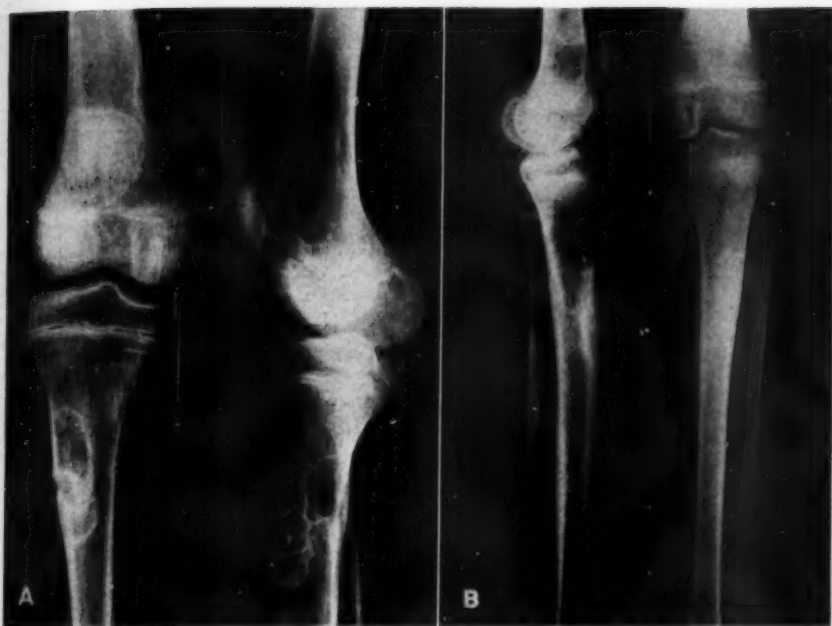


Fig. 14. Case 3: A. Two "cyst"-like areas in shaft of tibia and smaller area in femur from fibrous dysplasia of bone. No other involvement was found at this time. Areas were excised. B. Complete healing and filling in of resected area with new bone formation. No recurrence.



Fig. 15. Case 3: Unusual areas of pigmentation on the back over the spine.



Fig. 16. Case 3: Further development of polyostotic fibrous dysplasia in opposite radius a number of months later.

CASE 3 (Figs. 14-16): J. W., white female, age 10 years, was admitted to St. Louis City Hospital for probable injury to the right leg. X-ray examination showed two circinate cyst-like areas in the up-

per third of the shaft of the right tibia with beginning lesions in the lower part of the femur. No other



Fig. 17. Case 4: Large triangular area of pigmentation on under side of thigh, near vulva.

involvement was found at this time. The involved areas in the tibia were excised, without evidence of local recurrence, and were submitted for microscopic examination. These areas rapidly filled in with healthy new bone formation. Subsequent examination, however, revealed development of typical bony lesions in the lower end of the opposite radius.

This patient showed several small pigmented areas in the skin over the back, in the thoracolumbar region. There was early enlargement of the breasts, with development of pubic hair; menstruation began at eleven years of age.

Blood Examination: Red cells 4,440,000; 13.0 gm. hemoglobin; white cells 11,200; blood calcium 10.0 mg. per 100 c.c.; phosphorus 5.8 mg. per 100 c.c.; alkaline phosphatase 8.2 Bodansky units; Kahn test, negative.

No new areas of involvement have appeared.

CASE 4 (Figs. 17-20): E. G., white female, age 17 years, entered St. Louis City Hospital for emergency roentgen examination of the skull following an accident. No evidence of fracture was found but extensive cyst-like involvement of the skull was shown, with thickening of the outer table and sclerosis, giving an appearance similar to Paget's disease. Examination of the rest of the skeleton revealed involvement of the shaft of the left tibia, with rounded cyst-like areas characteristic of fibrous dysplasia of bone. The right humerus showed similar changes and in numerous ribs there were expanded areas of involvement. Microscopic examination revealed typical fibrous dysplasia of bone.

There was a large triangular area of pigmentation high up on the inner side of the right thigh. Enlargement of the breasts, pubic hair, and menstruation had appeared at nine years of age. The patient had been large for her age and had difficulty obtaining entrance to theaters, etc., at children's rates. At the age of nine she was 5 feet tall and "stopped growing at that time." At an early stage, a local physician requested her to enter a hospital for study.

Blood Examination: Red cells 3,780,000; hemoglobin 10.0 gm.; white cells from 7,950 to 18,000 (pelvic inflammatory disease); Kahn test, negative; acid phosphatase 0.8 and alkaline phosphatase 7.1 Bodansky units; blood calcium 9.5 mg. per 100 c.c.; blood phosphorus 3.5 mg. per 100 c.c.

This case of polyostotic fibrous dysplasia is remarkable for its extensive "cyst"-like involvement of the calvarium, closely resembling Paget's disease.

SUMMARY

Polyostotic fibrous dysplasia of bone as a part of Albright's syndrome (pigmented

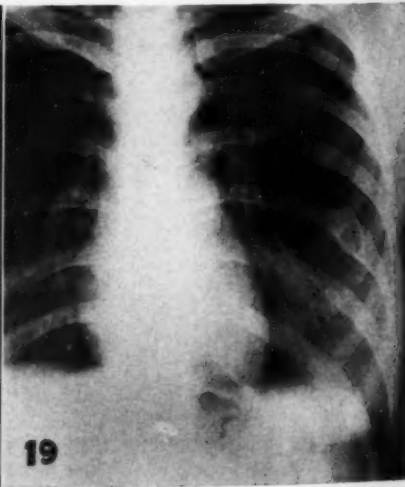
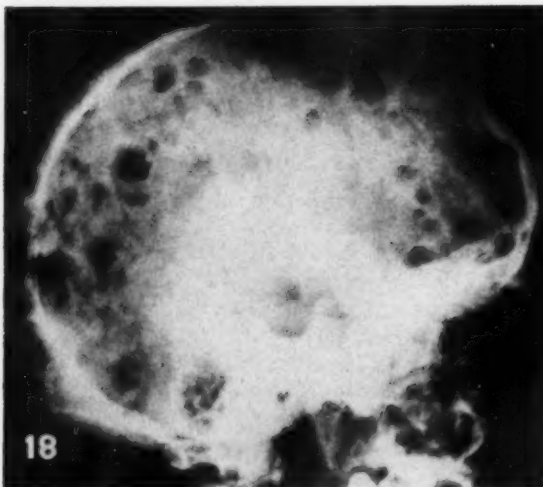


Fig. 18. Case 4: Extensive cystic development in calvarium of skull in case of polyostotic fibrous dysplasia.
Fig. 19. Case 4: Expanding lesions on ribs due to polyostotic fibrous dysplasia.



Fig. 20. Case 4: Other lesions of fibrous dysplasia in long bones.

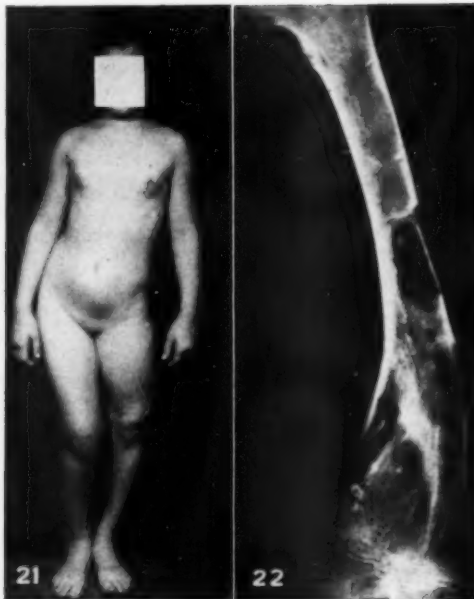


Fig. 21. Dyschondroplasia. Note deformity and shortening of femur.

Fig. 22. Dyschondroplasia (Ollier's disease), showing characteristic appearance of bone lesions at diaphyseal end of bone with areas of intervening decrease in density due to remnants of cartilage.

Irregular growth results in deformity and shortening of the bone.

chondroplasia (Ollier's disease) is illustrated.

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areas of the skin and precocious sexual development in the female) has been established as a definite entity. It undoubtedly occurs much more frequently than formerly supposed.

Its roentgenologic, pathologic and clinical features have been described and correlated.

Differentiation from "cyst"-like lesions produced in generalized von Recklinghausen's disease (osteitis fibrosa cystica) and other bone diseases has been discussed.

The essential variations between polyostotic fibrous dysplasia and dyschondroplasia (Ollier's disease) have been cited.

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SUMARIO

La Displasia Fibrosa Osteótica (Síndrome de Albright) y su Comparación con la Discondroplasia (Enfermedad de Ollier). Correlación de los Hallazgos Patológicos y Radiológicos

La displasia fibrosa poliosteótica como parte del síndrome de Albright, que también comprende pigmentación cutánea y, en la mujer, desarrollo sexual precoz, está ya establecida como entidad bien definida.

Clínicamente, constituye un proceso lento e incoercible, que puede hallarse ya bien desarrollado en todos los tejidos óseos del cuerpo antes de que se sospeche su presencia debido a disfunción o fractura patológica. El diagnóstico suele ser radiográfico.

En el diagnóstico, las lesiones más importantes, aunque no absolutamente características, son las zonas quistoideas redondeadas de expansión unilateral dentro de la corteza de las diáfisis de los huesos largos. Obsérvase adelgazamiento en cascarrón de la corteza, que ocasiona fragilidad ósea y fracturas espontáneas, las cuales cicatrizan fácilmente por neostogenia. No se notan elevación perióstica, salvo por reacción a fracturas, ni osteoporosis ni deformidades incurvadas de los huesos. Puede sobrevenir alguna esclerosis, si se afectan los huesos menos cancelosos. Preséntanse ensanchamiento y alargamiento de las diáfisis. La tendencia es a invasión unilateral.

El examen microscópico revela absorción osteoclástica del tejido óseo, seguida de fibrosis de la médula ósea en las zonas corticales y subcorticales. El proceso comienza en los conductos de Havers y se difunde a los tejidos circundantes. La

corteza se adelgaza y vuelve trabecular, reabsorbiéndose muchas trabéculas de la porción esponjosa del hueso, en tanto que llena los espacios un tejido fibroso más o menos flojo.

En la displasia fibrosa poliosteótica la invasión ósea semeja en algunos sentidos la osteítis fibrosa quística (enfermedad de von Recklinghausen), pero discrepa de ésta en la falta de osteoporosis. Además, las partes dilatadas en las zonas corticales y subcorticales no son verdaderos quistes, que contienen líquido, sino que están llenas de tejido fibroso. También hay que diferenciar el estado, de la discondroplasia o enfermedad de Ollier, constituyendo características diferenciales el origen de la última en el tejido canceloso de la región de las metáfisis y la aparición de las lesiones de la displasia fibrosa en las regiones corticales de las diáfisis de los huesos largos. Roentgenológicamente, la enfermedad de Ollier muestra hebras longitudinales de las trabéculas óseas conservadas en forma de abanico en las epífisis de los huesos afectados, con las zonas enrarecidas ocasionadas por los depósitos cartilaginosos interpuestos. En esas zonas moteadas descúbrese cuerpecillos globulares intensamente calcificados. Patológicamente, las zonas enrarecidas proceden, según se demuestra, de la proliferación cartilaginosa en el tejido óseo que ocasiona disosteogénia, y no de los depósitos de tejido fibroso o de quistes serosos.

Occult Meningocele of the Sacrum

Report of Three Cases¹

V. W. ARCHER, M.D., GEORGE COOPER, JR., M.D., and C. V. CIMMINO, M.D.

THE TERMS "occult meningocele" and "spina bifida occulta" have occasioned confusion in the past. Kaufmann implies that these are more or less synonymous. Adami, however, considers "meningocele" to be a cyst of the meninges, or a hernia, projecting through the walls of the vertebral canal, and "spina bifida occulta" to be a localized lack of junction of the laminae with no projecting fluid sac.

In this paper the term "occult meningocele" is employed rather than the often used "spina bifida occulta." It embraces a diffuse dilatation of the dural sac with or without the relatively inconsequential element of herniation into the soft tissues. The meningocele, rather than the bifid spinous process, is the significant element. In one of the three cases upon which this report is based there was no spina bifida.

Mixer (1), in his section in Lewis' "Practice of Surgery," gives an excellent description of this condition, using the term "spina bifida occulta." He describes it as a congenital malformation consisting of a defect in the closure of the vertebral arch, usually with an associated defect of the meninges and nerve tissues. He also states that, although there is no external meningeal sac, there is frequently a localized growth of hair. This was not present in any of our three cases. Mixer also adds that there is almost invariably a congenital lipoma either just beneath the skin or in some of the deeper structures. This was not true of our single operative case.

In this same article the anatomical and pathological basis for the usual symptomatology is presented, and to this presentation the reader is referred for greater detail. In brief, Mixer describes one defect which is almost constant, the fusion of the central portion of the cauda equina into a

single irregular bundle, which may be the site of various pathological processes comprising the nerve roots and causing disturbance of function. He mentions lipomata, dermoid cysts, and a band of dense connective tissue.

Mixer also states that there are two types of history: one in which paralysis has been present since birth and one in which symptoms came on in later life. In one of our cases, the symptoms were precipitated in middle life by an injury which evidently affected the thin bony shell over the sac (Case III). The usual striking symptom is lack of bladder control, present in two of our cases, absent in the third. Occasionally, there are toe drop and a saddle anesthesia.

Enderle (2), in 1932, demonstrated an occult intrasacral meningocele by myelography, which he thought was the first to be described in the roentgen literature. The patient was a forty-seven-year-old woman with pain radiating down the posterior surfaces of both legs. Plain films showed a large sacral hiatus and lumbarization of S-1. A normal cul-de-sac was demonstrated on immediate examination. Twenty-four hours later the oil descended into a lower sac, $7 \times 4 \times 2$ cm., which was united to the lumbar sac by a narrow isthmus.

Wilson and Wakeley (3), also in 1932, described a case of occult meningocele with no symptoms until the age of sixteen. At that time, pain occurred in both legs, and later bladder and rectal incontinence developed. At the time of observation, two years after onset of symptoms, pain had become excruciating. Roentgen examination showed a circular defect in the back of the sacrum and fifth lumbar vertebra. At operation, there was no evidence of the

¹ From the University of Virginia Hospital and Medical School, Charlottesville, Va. Presented at the Thirty-third Annual Meeting of the Radiological Society of North America, Boston, Mass., Nov. 30-Dec. 5, 1947.

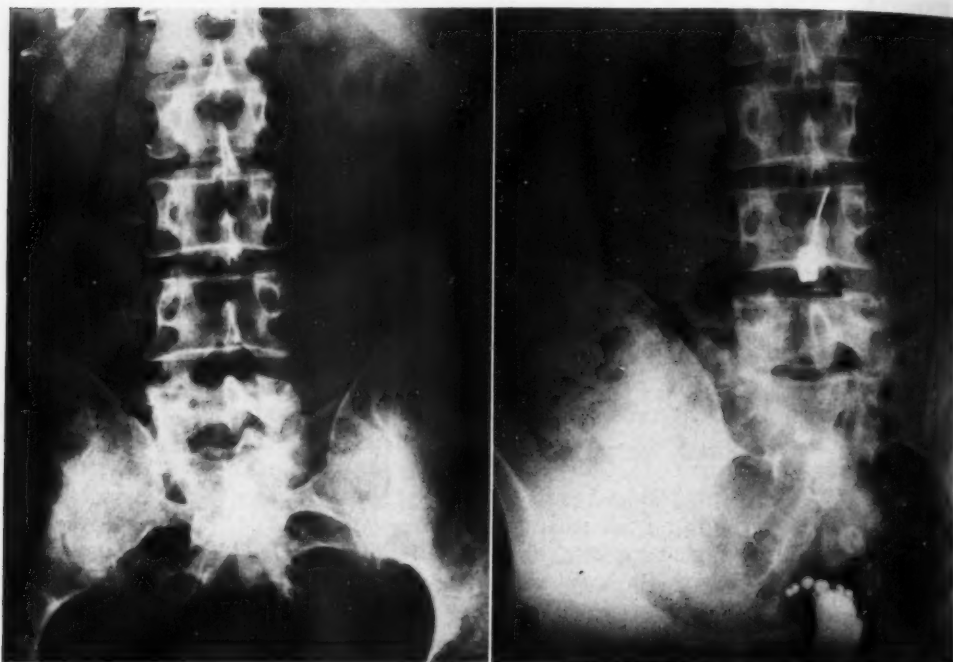


Fig. 1. Case I: Note defect in the vertebral arches of the sacrum and the large accumulation of lipiodol far below the usual caudal limit of the dural sac.

presence of laminae or spinous processes of the fifth lumbar vertebra, but a meningeal hernia was encountered in the soft tissues. There were no external stigmata such as localized tufts of hair or a dimple to suggest the presence of this lesion, and no prominence over this portion of the spine.

The histories of our three patients are as follows:

CASE I: J. K. H., a 43-year-old white man, gave a history of aching pain in the right buttock for four years. The pain gradually spread to the right thigh and to the back of the right ankle, which became weak. On admission, a severe degree of right pes cavus was found, absence of the right ankle jerk, atrophy of the right calf, weakness of the right hip flexor muscles, and hypesthesia over the right first sacral dermatome.

Roentgen studies (Fig. 1) revealed a probable sacral meningocele, but the patient was discharged without surgery.

CASE II: E. H., a 38-year-old white man, stated that he had injured his sacrum at the age of nineteen, since which time he had suffered from sacrococcygeal pain. A year later, he began to lose con-

trol of the urinary sphincter, had trouble starting the urinary stream, and suffered from blinding headaches, blurred vision, syncope, and sensory disturbances over the sacrococcygeal area and thighs. At the age of thirty-three, pyonephrosis necessitated a right nephrectomy, followed shortly by a suprapubic cystostomy because of poor bladder function. The urinary status has never been good enough to consider surgical investigation of a probable sacral meningocele (Fig. 2A). Roentgen studies showed a block in the sacral canal (Fig. 2B).

CASE III: R. A. C., a 40-year-old white woman, complained of severe sacrococcygeal pain after a fall five months before admission. There were fullness and tenderness over the sacrum just to the left of the mid-line. Roentgen studies (Fig. 3) led to a presumptive diagnosis of neurofibroma. At operation, the normal sacral landmarks were obliterated and the bone presented a smooth convex surface. A meningocele the size of a lemon was discovered beneath this paper-thin cortex of bone. The defect was repaired. Because of recurrence of the hernia, a tantalum plate was inserted into the defect several weeks later. Herniation still persisted, and after five weeks a complete repair was finally effected, although low back pain persisted. Two disk operations followed, and finally, about two years ago,



Fig. 2. Case II: A. Cystic defect in lower sacrum. B. Lipiodol in sacral sac.

the left sensory roots of the fifth lumbar and first sacral nerves were sectioned. The back has been relatively asymptomatic to date.

DISCUSSION

These three patients presented widely differing symptoms, one gradually increasing pain, one pain following trauma, and the other the usual bladder symptoms of incontinence. Two of these patients were forty years old when symptoms first appeared, the third twenty years of age. None of the three presented external stigmata suggesting the underlying process. A definite meningeal hernia was encountered in the sole operative case (Case III).

It is suggested, therefore, that in all patients with symptoms which might be referable to the lumbosacral region, particular attention be paid to possible enlargement of the canal within the sacrum. In our limited experience with this lesion, we have come to the conclusion that in all patients with these complaints, it might be well to make additional lateral views of the sacrum, using soft-tissue technic. One of our cases (Case III) certainly would have

been overlooked otherwise, since there was an eggshell posterior plate, barely visible on a very soft film.

DIFFERENTIAL DIAGNOSIS

Since the pathological process is an enlarged lower dural sac, any lesion, such as neoplasm, producing a relatively radio-translucent zone within the sacrum may simulate meningocele. Camp and Good (4) give a comprehensive discussion of sacral neoplasms, dividing them into three groups:

1. Tumors arising within the sacral canal, of which the ependymal-cell glioma is the commonest. These cause a smooth erosion of bone which may be uni- or multiloculated, with decalcification of the surrounding bone. Decalcification of the neural arches due to bone erosion is usually absent in occult meningocele.

2. Tumors arising within the sacrum proper, of which the chordoma is commonest. The malignant tumors, including the occasional cases of Ewing's tumor and osteogenic sarcoma, present the usual characteristics of malignant bone lesions and

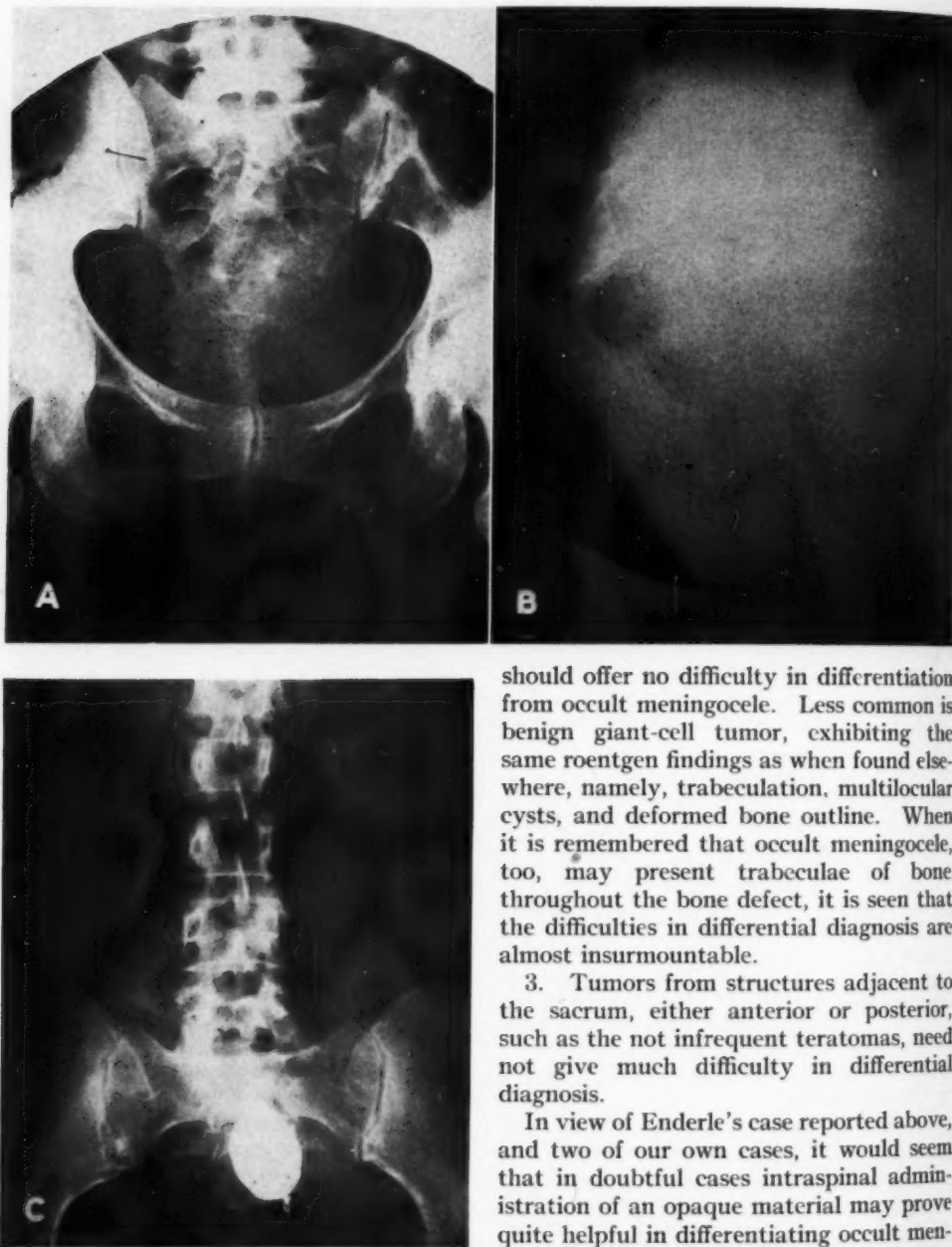


Fig. 3. Case III: The original films, from which a diagnosis of neurofibroma was made, showed a thin plate of bone over the dorsum of the sacrum. This is visible only in the lateral view, with soft-tissue technic (B), and does not reproduce well. The tantalum plate inserted following a postoperative recurrence is also illustrated (C).

should offer no difficulty in differentiation from occult meningocele. Less common is benign giant-cell tumor, exhibiting the same roentgen findings as when found elsewhere, namely, trabeculation, multilocular cysts, and deformed bone outline. When it is remembered that occult meningocele, too, may present trabeculae of bone throughout the bone defect, it is seen that the difficulties in differential diagnosis are almost insurmountable.

3. Tumors from structures adjacent to the sacrum, either anterior or posterior, such as the not infrequent teratomas, need not give much difficulty in differential diagnosis.

In view of Enderle's case reported above, and two of our own cases, it would seem that in doubtful cases intraspinal administration of an opaque material may prove quite helpful in differentiating occult meningocele from tumor. The demonstration of a large sac into which oil flows from a higher level is conclusive proof of the nature of the cavity which has been demonstrated.

SUMMARY

1. Three cases of occult meningocele of the sacrum are reported.
2. The symptomatology may be pain, loss of reflexes, saddle anesthesia, or incontinence.
3. External stigmata are not invariably present.
4. Additional soft-tissue lateral films are recommended in suspicious cases.
5. Spina bifida is not invariably present.
6. Intraspinal injection of an opaque medium may be helpful in differential diagnosis between occult meningocele and intrasacral tumor. In view of Enderle's

case, a twenty-four hour film is recommended.

7. It is suggested that the term "occult meningocele" be used instead of "spina bifida occulta."

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SUMARIO

Meningocele Oculito del Sacro. Tres Observaciones

Comunicanse tres casos de meningocele oculito del sacro, prefiriéndose esta designación a la de espina bífida oculita, por ser el meningocele el elemento importante.

Los síntomas del meningocele oculito comprenden dolor, arreflexia, anestesia en silla de montar e incontinencia urinaria. En ninguno de los casos aquí descritos había presentes estigmas externos.

En los casos sospechosos recomiéndanse radiografías laterales de los tejidos blandos. Para el diagnóstico diferencial con tumor intratecal, puede ser útil la inyección intrarraquídea de un medio opaco. Recomiéndase una película de 24 horas. El hallazgo de una bolsa grande en la cual mana el aceite desde más arriba establece el diagnóstico.

DISCUSSION

(Papers by Heublein, Bernstein, and Hubenet; Holt and Wright; Ball and Legant; Sante, Bauer, and O'Brien; Archer, Cooper, and Cimmino)

Joseph Barr, M.D. (Boston, Mass.): All orthopedic surgeons lean heavily upon the radiologist and his field in orthopedic work, whether it be with children or adults, and I should like here briefly to pay tribute to my teachers in that field in past years—Dr. Sosman at the Peter Bent Brigham Hospital, Dr. Vogt at Children's, Dr. Holmes, Dr. Hampton, and others at Massachusetts General, who taught me what little I know about x-ray diagnosis.

It would be impossible for me to discuss in any sensible sort of fashion all the things that have been brought before you this afternoon. I shall therefore mention just one or two points from each paper which interested me.

Ectopic bone formation about joints, of which Dr. Heublein showed one example, is certainly a rare condition. I have seen one similar case which was a post-encephalitic manifestation. The

child had measles, then encephalitis, was unconscious for some time, developed ankylosis of the elbows and hips, and showed roentgenographically an appearance very similar to that which we saw in Dr. Heublein's films—bridges of bone which extended just outside the capsule of the joint. I operated on one of the elbows and by removing this bridge of bone was able immediately, on the operating table, to restore a completely normal range of motion to the elbow. The articular cartilage remained intact. These were extracapsular ossifications.

Congenital coxa vara is an interesting entity which anyone doing much children's radiology will occasionally see. It should be recognized as a congenital malformation in which ossification does not take place satisfactorily in the cartilaginous matrix. The result is coxa vara with improper ossification of the neck.

I was much impressed by the paper of Dr. Holt and Dr. Wright on fibromatosis, a copy of which they had previously forwarded to me. This was extraordinarily complete, and the essayists were not able to present today nearly all the factors which they took up in the paper. Two things which they mentioned are of great interest to orthopedists. One is the matter of scoliosis. When you see a film which shows scoliosis, there should be a careful search for neurofibromatosis or osseous or juxta-osseous lesions. This type of scoliosis is frequently resistant to therapy and we should make the diagnosis, as it will help us in prognosis and in treatment. It is practically impossible, I believe, to correct these curves satisfactorily.

Congenital pseudo-arthritis is another extremely difficult orthopedic lesion to treat, and in some instances we must resort to amputation.

It seems to me that radiologists should put pressure on hospitals to have skeletal surveys of patients. Such surveys could be carried out without great cost if suitable methods were devised. We require routine chest films in many hospitals today. I suspect that the time isn't too far distant when we will require routine skeletal surveys.

I know nothing about sickle-cell anemia. We have a small Negro population and I have seen no such cases. The osseous lesions are certainly like caisson disease, of which I have seen a moderate amount. I wonder, if we surveyed the skeletal systems of patients with other thrombotic diseases, whether we might not find similar lesions. The malignant type of malaria might produce a similar osseous picture.

I wonder if perhaps the use of anticoagulants in sickle-cell anemia might not be worth while. I have seen one case of Cooley's anemia recently, in an adult, with an appearance not at all like that shown in sickle-cell anemia. In Cooley's anemia there is a disappearance of the osseous cortex with an extreme ground-glass appearance of the bone.

Polyostotic fibrous dysplasia, as described by Sante, Bauer, and O'Brien, is a very interesting entity. I don't personally think that it looks very much like Ollier's disease and suspect that in most cases the differential diagnosis can be made even without pathological section.

We finally come to the tail end of the program. I wonder if they put this last paper there purposely. Sacral meningocele is a rare lesion, certainly. I have seen only one case but it adds just one more diagnosis to the problem of back pain, and we must be on the lookout for it as a rare cause of low back pain and of radiating pain.

Stanley H. Macht, M.D. (Baltimore, Md.): For the past year, down in Baltimore, Dr. Paul Roman and I have been studying all the sickle-cell anemia cases which have been seen there since 1936. Out of 70 cases, we finally derived 48

which were proved true sickle-cell anemia.¹ Our findings were quite parallel to those described by Doctors Ball and Legant.

In 42 of our 48 cases, chest films were obtained; 75 per cent showed cardiac enlargement and 38 per cent of these 42 cases showed coincident consolidation of some portion of the lung.

The skull changes were not significant. Only 8 cases showed changes in the bones of the skull. All of these changes were noted in children. They did not present the so-called typical or classical textbook "hair-on-end" appearance, but showed a stippling due to hyperplasia of the erythroblastic tissue.

I should like to show two slides illustrating this hyperplasia. [Slides were shown at this point.] One is a normal specimen of bone marrow. This I think forms a rather striking contrast to sickle-cell bone marrow, which is shown on the next slide. Here you see the absence of fatty elements, the hyperplastic marrow, and the atrophic bony spicules which stand out on an x-ray film. These changes probably account for the osteoporotic appearance that Dr. Legant described.

There is one other thing that I think should be emphasized in this discussion, and that is the type of reaction which one sees in the bones of a child's hand. We have three cases in which the bone lesion involved the metacarpals. No bone change was observed on the initial study, even though there was a clinical picture of heat, swelling, and pain; but after a period of three weeks, periostitis and necrosis occurred and could be visualized roentgenographically. These are only transient and may be missed entirely if one does not bring the patient back for re-examination three or four weeks after the onset of clinical symptoms. The time element is an important factor in these changes and should be stressed.

Dr. Heublein (closing): I think we saw a great many examples of myositis ossificans during World War II due to high explosive fragments. We have discussed these with our orthopedic and Army doctors and have come to the conclusion that they were due to a combination of infection due to large decubitus ulcers in the region of the trochanters, transverse myelitis, and a long period of dorsal recumbency.

Dr. Legant (closing): I will be very brief. The same thought occurred to us as to Dr. Barr. Why shouldn't we see bone infarcts with other blood dyscrasias in which a thrombotic tendency exists? The obvious one is polycythemia vera. We have found a bone infarct in the humerus of a patient with long-standing polycythemia vera. It was in no way different from the infarct in the humerus which we saw in caisson disease.

¹ See paper by Macht and Roman page 697 of this issue.

The Radiologic Changes in Sickle-Cell Anemia¹

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SINCE HERRICK (1) described sickle-cell anemia in 1910 there have been many reports of this disease in the literature. These have dealt with studies of the cell, the pathological basis of the disease, and the common as well as uncommon clinical findings. It is the purpose of this paper to present a radiologic study of sickle-cell anemia in order to demonstrate the effects on the entire organism, rather than on the bones alone, which can be recognized by radiological examination.

Sickle-cell anemia affects all tissues of the body. Although there is a characteristic clinical picture, there are many variations depending on the particular pathological involvement of various organs, and because of this Wintrobe (2) has aptly named the disease "the little imitator." Many investigators have discussed the theories of the cause of sickling. These have dealt with heredity, oxygen tension studies, and immunologic factors (3, 4, 5).

The sickled cell loses its flexibility and appears fixed and rigid. Its length may be two to five, or more, times its original diameter, with long tapering processes projecting from the ends. Murphy and Shapiro (6) show that "capillary blockades" can be formed by these sickled cells. These lead to stasis of blood in the capillaries and small blood vessels, thromboses, and infarction. The clinical picture is associated with these thromboses and infarctions. The changes in the bones are due to two factors: (a) hyperplasia of the marrow secondary to a severe degree of hemolysis; (b) congestion and thrombosis, infarction, scarring, and regeneration of bone (6-13).

The nervous system may be affected (14-18). Klinefelter (19), McKendry (20), Zimmerman and Barnett (21), Winsor and

Burch (22), and Wintrobe (23) have shown the heart to be frequently involved. Indolent skin ulcers and other skin changes are often seen (24-26). Cholelithiasis and cholecystitis in young patients have been found by Schaefer (27) and Weens (28). This is thought to be due to the hyperbilirubinemia resulting from increased hemoglobin liberation.

Progressive renal damage with fixation of the specific gravity is a frequent finding. Bauer (29), Diggs and Ching (30) and Yater and Hansmann (31) have shown this to be associated with a pathological picture of destruction of the glomeruli by accumulations of sickled red blood cells. Pulmonary thrombosis and infarction are common (30-31). Getzoff (33) reports priapism as a result of thrombosis. The pathological findings in the liver and spleen are well known (6, 7, 11, 30, 32).

The factors that are responsible for the production of the crises of sickle-cell anemia are not well understood. Sherman (5) has found by experiments that increased leukocyte concentration, temperature elevation, and bacterial contamination are associated with acceleration of sickling. Clinically, infection is supposed to precipitate the sickle-cell crisis (2). The theory has been advanced that the maturing red blood cells with their increased tendency to sickle, in association with increased coagulability of the blood, suddenly form a massive thrombus, and this results in a crisis (6). The clinical manifestations of sickle-cell anemia will depend, therefore, upon the effects produced by a chronic hemolytic anemia and by thromboses and infarction of one or more organs. They will also depend on the efforts of the body to overcome and heal the effects of anemia and thrombosis.

¹From the Department of Radiology, Baltimore City Hospitals. Accepted for publication in December 1947.

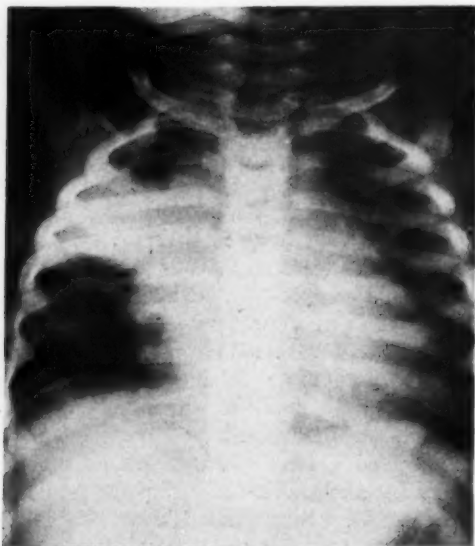


Fig. 1. Consolidation of a segment of the lung associated with cardiac enlargement. Pneumonia is frequently a precipitating factor of sickle-cell crises. In addition to the straight cardiac border seen above, the pulmonary conus is often very prominent, producing the picture seen in mitral heart disease.

In considering the roentgenologic aspects of sickle-cell anemia it was felt it would be of interest to determine the changes demonstrable by roentgen study of the skeleton, chest, and abdomen. A search of the literature fails to reveal a classification of the radiographic changes

in this disease. Based on a study of the cases seen in this series, the following classification has been derived:

- I. Radiographic Evidence of Involvement of the Viscera
 - (a) Cardiac enlargement
 - (b) Pathological changes in the lungs
 - (c) Enlargement of the liver; cholelithiasis
 - (d) Enlargement of the spleen in children—possible calcifications later in life
- II. Skeletal Changes Due to Hyperplasia of the Erythroblastic Elements
 - (a) Long bones
 1. Irregular trabeculation
 2. Widening of the medullary spaces
 3. Cortical thinning
 - (b) Short bones and flat bones
 1. Exaggeration of trabecular markings of ilia, scapulae, phalanges
 2. Cupping of vertebral bodies
 3. Skull: Widening of diploe, thickening of parietal and frontal bones, thinning of outer tables, and radial arrangement of trabeculae
- III. Skeletal Changes Due to Thromboses
 - (a) Long bones
 1. Cortical thickening
 2. Narrowing of medullary cavity
 3. Loss of bone tissue
 4. Periosteal reaction
 - (b) Short bones
 1. Necrosis and resorption of bone
 2. Periosteal reaction
- IV. Skeletal Changes Due to Disturbances of Growth

TABLE I: ROENTGEN FINDINGS IN FORTY-EIGHT CASES OF SICKLE-CELL ANEMIA

Part X-Rayed	No. Cases Examined	Positive Findings	Cases with One or More Positive Findings
		Cases	
Chest	42	Cardiac enlargement.....32 (76.2%)	32 (76.2%)
		Pneumonia.....16 (38%)	
Long bones	35	Coarse trabeculation of ribs and scapulae. 3 (7.1%)	23 (65.7%)
		Hyperplasia of marrow.....20 (57.1%)	
		Growth disturbance.....8 (22.8%)	
		Periostitis.....6 (17.1%)	
		Bone destruction.....3 (8.6%)	
		Sclerosis of bone.....2 (5.7%)	
		Joint destruction.....2 (5.7%)	
Abdomen (pelvis and spine)	23	Splenomegaly.....8* (34.8%)	12 (56.5%)
		Hepatomegaly.....8 (34.8%)	
		Vertebral cupping.....4 (17.4%)	
		Cholelithiasis.....2 (8.7%)	
Short bones (hands and feet)	17	Coarse trabeculation.....6 (35.2%)	9 (52.8%)
		Periostitis.....3 (17.6%)	
		Destruction.....2 (11.7%)	
Skull	30	Prominent diploe.....7 (23.3%)	8 (25.9%)
		Radial striations.....2 (6.7%)	
		Thickening of bones.....1 (3.3%)	

* One case also showed amorphous calcifications in the enlarged spleen.

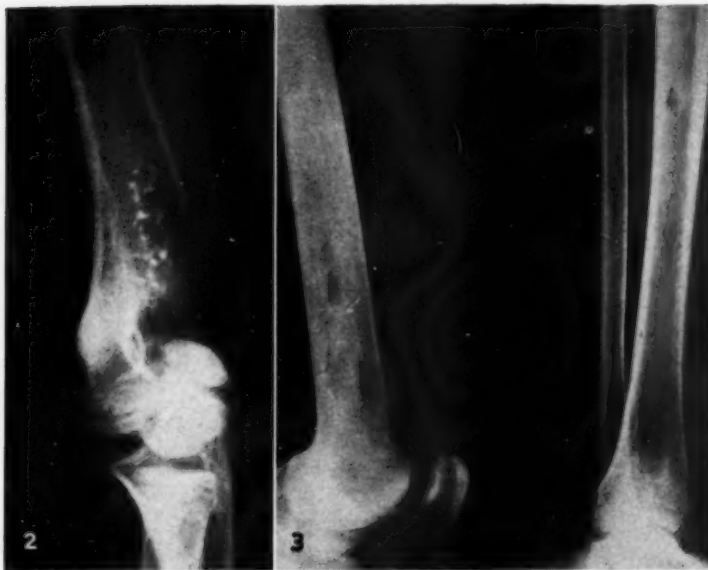


Fig. 2. Changes in the long bones are produced by hyperplasia of the bone marrow. Note coarse trabeculae, widened marrow cavity, irregularity of the inner border of the cortex and thinning of the cortex.

Fig. 3. Thromboses may produce areas of bone destruction. Note punched-out areas, as well as widening of the marrow cavity and thinning of the cortex of the fibula.

The present study is a review of 48 cases of proved sickle-cell anemia indexed in the records of the Baltimore City Hospitals during the years 1936 through 1947. Twenty-nine of the patients were children under fourteen years of age. All cases of sickle-cell trait (sickleemia) without anemia were excluded from this series. According to the present criteria for a complete roentgen examination of the patient with sickle-cell anemia, most of the cases were incompletely studied. Only those seen since this study was undertaken have received a comprehensive x-ray examination. Such an examination should include films of the skull, chest, abdomen and pelvis, long bones, hands and feet. The films for 19 of the cases had been discarded and in these instances the filed reports were carefully reviewed. Each of the reports had been made by an experienced radiologist.

An analysis of the findings in these 48 cases disclosed the data shown in Table I.

From the table it can be seen that involvement of the heart is the most fre-

quent finding, enlargement occurring in 76.2 per cent of 42 cases. This has also been described by Klinefelter and others (19-23). Both fluoroscopically and on films the heart appears diffusely enlarged (Fig. 1) and often has a prominent pulmonary conus, giving it the so-called "mitral configuration" seen in rheumatic heart disease.

Consolidation of various segments of the lung (Fig. 1) occurs frequently. Clinically the picture is one of a lobar or bronchopneumonia and not one of infarction. As the patient improves clinically, the abnormal roentgen findings in the lungs disappear. Quite often sickle-cell crises are precipitated by pneumonia. Of 42 cases in this series 38 per cent showed pneumonia.

One of the most frequent x-ray changes noted is a coarsening of the trabeculae of the long bones associated with widening of the marrow cavity and thinning of the cortex (Figs. 2 and 3). The probable explanation for this is hyperplasia of the

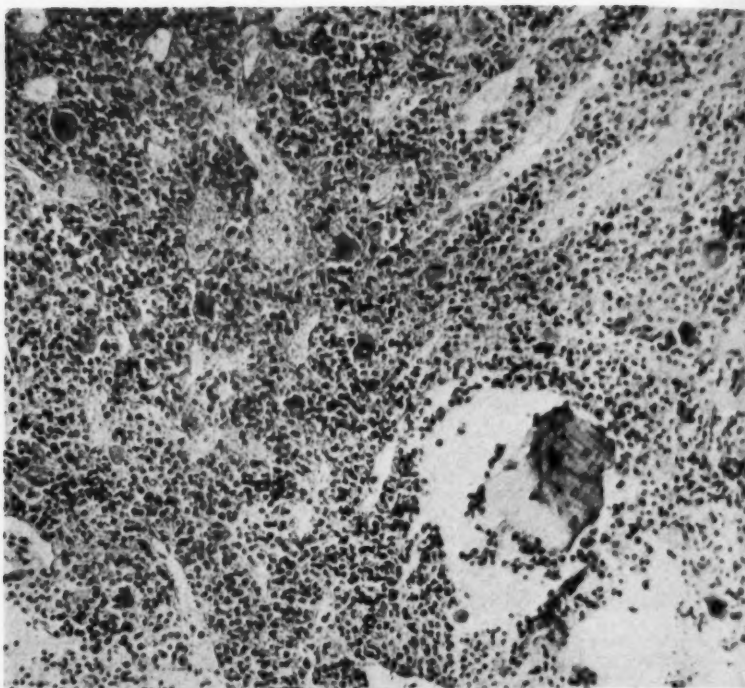


Fig. 4. Photomicrograph of section of bone from patient who died of sickle-cell anemia. Note hyperplastic packed erythroblastic tissue with absence of the usual lace-like appearance of marrow. An atrophic, demineralized bone spicule is seen in the right lower corner.



Fig. 5. Coarse trabeculae are also seen in the acromion processes and in the ribs. These may be detected on the chest film. When the disease has progressed enough to produce these changes, the heart will also be enlarged.

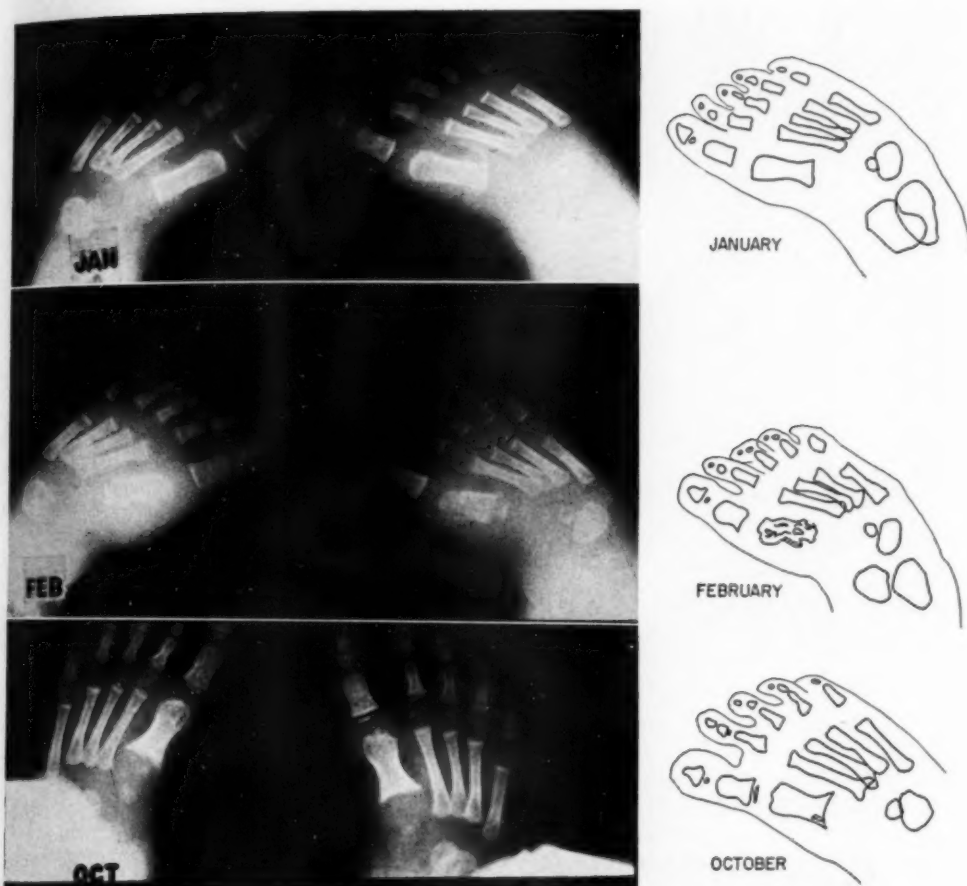


Fig. 6. Films of the feet made in January showed normal bone structure. During a crisis in February there was swelling of the hands and feet. Roentgen study at this time showed destruction of each first metatarsal. The process gradually subsided during the succeeding months until, by October, the bones were completely regenerated. The drawings illustrate the changes shown in the films.

marrow elements. These press upon the bone spicules and produce atrophy (Fig. 4). The spicules which remain in the widened marrow cavity are therefore more conspicuous on the x-ray film, standing out in contrast because of the absence of other trabeculae which have been destroyed. The cortex of the bone becomes thinned and atrophic by the pressure of the packed-in hyperplastic marrow. These changes have been described by Swenson and Caffey (34).

In the flat bones and short bones this same process occurs but to a lesser degree because of the relative decrease in the

erythroblastic tissues in these bones. Coarse trabeculae were seen in the short bones of the hands and feet in 35.2 per cent of 17 cases and less often in the ribs and scapulae (Fig. 5).

Thromboses may occlude the capillaries (11) of the bones. When this occurs, there are infarction, necrosis, and resorption of the bone (Figs. 3 and 6). After healing occurs the picture is similar to that seen in caisson disease (Figs. 7 and 8).

It has been our observation that a definite time interval occurs between the clinical signs and symptoms of swelling, heat, and pain of the hands or feet and the



Fig. 7. Bone infarction with subsequent repair produces areas of sclerosis similar to the changes noted in caisson disease.

Fig. 8. Thromboses may damage the articular ends of bones and eventually destroy a joint. The above changes may be mistaken for those of "coxa malum senilis" so often seen in the aged. However, this is a film of the hip of a 27-year-old colored male who has had frequent sickle-cell crises since childhood, and multiple bone lesions.

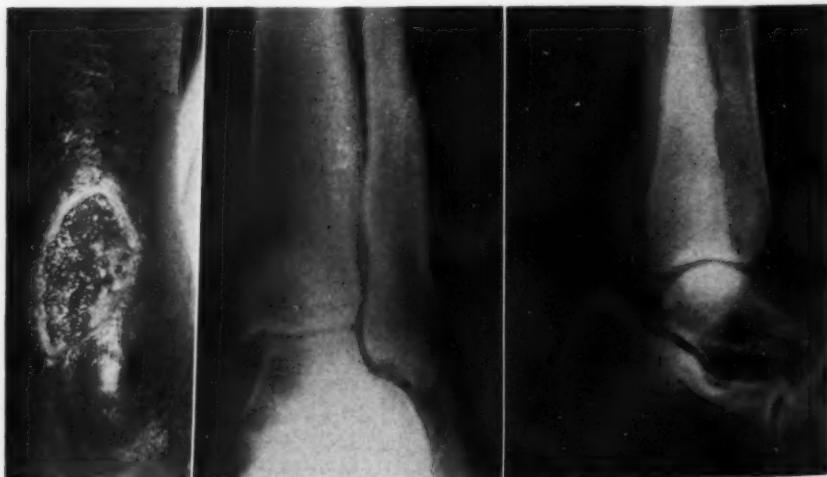


Fig. 9. Chronic ulcers, most commonly seen about the ankle, produce changes in the underlying bones. Periosteal thickening and periostitis produce a bone picture identical with that seen under long-standing varicose ulcers.



Fig. 10. A. Periosteal thickening along the shaft of a long bone. Such thickening, when present, may be indistinguishable from that noted in leukemic bone involvement.

B. and C. Periosteal thickening due to leukemia (from Kalayjian *et al.*: *Radiology* 47: 229, 1946).

appearance of bone changes roentgenologically. This fact has not been previously emphasized in the literature. In two cases, signs and symptoms were noted and the initial roentgen findings were negative. In each case, however, films taken an average of twenty-one days later showed evidence of bone destruction involving both the marrow and the cortex. Repair occurs after a long period of time (Figs. 6, and 7). Danford *et al.* (13) describe a similar case in which recovery was more rapid, all bone changes having subsided by the forty-third day.

If the thromboses involve the blood supply near the articular end of a bone, there may be permanent destruction of the joint (Fig. 8). De Lorimier (35) presents such a case in his book on arthropathies. Two such cases, one in which there was destruction of the wrist and a second in which there was destruction of the hip, were observed in this series.

Thromboses of the capillaries of the skin,

especially common at the ankles, produce chronic ulcers which may not heal for years (Fig. 9). Long-standing infection and stasis are reflected in periosteal changes in the underlying bones. These latter changes are indistinguishable from those noted in the bones under long-standing varicose ulcers. Periosteal thickening may develop around the long bones and present a picture similar to that often seen in children with leukemia (Figs. 10A and C).

Generalized growth disturbance has been described and explained on a basis of poor nutrition of body tissues and endocrine glands as a result of the severe anemia. A definite "sickle-cell habitus" for patients with this disease has been demonstrated (36-38). Some authors believe the patient may show specific physical characteristics. The changes are secondary to the disease and consist of a linear habitus, emaciation, long extremities, short trunk, upper dorsal kyphosis, deep chest, protuberant abdomen, spider hands, and occasionally an

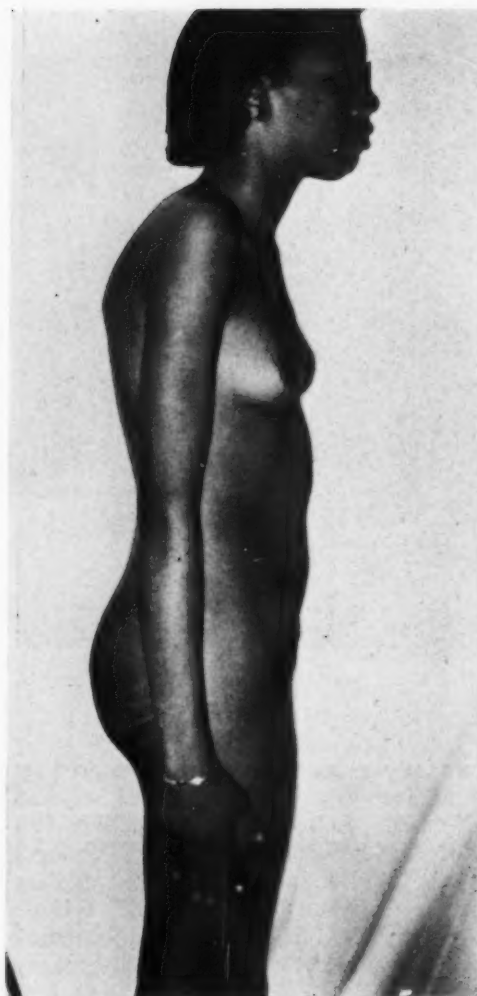


Fig. 11. This 23-year-old colored female presented many of the features characteristic of the "sickle-cell habitus." Note dorsal kyphosis, linear habitus and slight turriccephaly.

abnormally shaped head (Fig. 11). Abnormal dentition and delayed osseous union have also been found by Sharp and Vonder Heide (38). These findings are reflected in x-ray studies of these patients. The soft tissues of the extremities are thin and atrophic. Evidence of periods of severe growth disturbance is a common finding on the films (Fig. 12). Multiple bands of increased density near the epiphyseal ends of the long bones indicate periods of illness

when growth had temporarily stopped. Films of the spine show the dorsal kyphosis which is seen on physical examination. The weakening of the bone structure by atrophy of the spicules and by osteoporosis may explain this kyphosis. The transmitted pressure of weight through the nuclei pulposi on the bodies of the weakened vertebrae may cause them to become cupped on their superior and inferior surfaces (Figs. 13 and 14). This change has also been noted by de Lorimier (35), Winsor and Burch (22), and by Diggs, Pulliam and King (9).

A film of the abdomen of the patient with sickle-cell anemia may demonstrate one or more of the following: enlargement of the liver and spleen, cholelithiasis, and vertebral changes. Hepatomegaly and splenomegaly are more commonly seen in children. The enlargement of the spleen may occasionally be detected during the course of a barium enema study by observing an extrinsic pressure defect in the colon in the splenic area. Later in life there may be soft miliary-like deposits of amorphous calcium in the spleen. These can be seen on the x-ray film but are not "contrasty" enough to be seen in photographic reproductions. One case in this series showed these calcifications. The calcium is deposited in the hemoglobin-packed intersinusoidal pulp of the spleen after the breakdown of the conglomerated sickled cells. Gallbladder calculi may often be seen in young adults (Fig. 14).

The turriccephalic appearance of the skull is produced by an actual increase in the width of the parietal bones. Although most textbooks describe the "hair-on-end" appearance as characteristic of sickle-cell anemia, this change is only rarely seen. When changes occur, the usual ones are widening and prominence of the diploe. They produce a fine stippled appearance best seen in the frontal and parietal bones. The more advanced cases may show radial striations (Fig. 15). Of 30 cases in this series, 74.1 per cent showed no change.

Many authorities do not feel there are any specific changes characteristic of sickle-



Fig. 12. Multiple bands of increased density near the epiphyseal ends of the long bones indicate periods of severe illness, when the growth process was delayed.

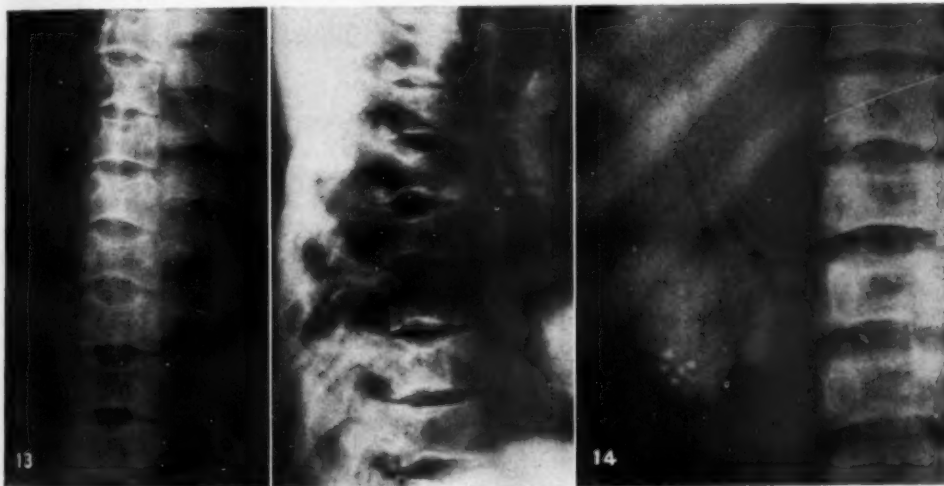


Fig. 13. Cupping of the superior and inferior surfaces of the vertebral bodies.

Fig. 14. Cholelithiasis, probably on the basis of hyperbilirubinemia. Cupping of the vertebrae is also shown.

cell anemia. The authors believe that the findings, while not characteristic, are sufficiently prominent for the radiologist to suggest the diagnosis and thus call it to the clinician's attention. Often to think of sickle-cell anemia is to diagnose it.

SUMMARY

1. Based on a radiologic study of 48

cases of proved sickle-cell anemia, a classification has been derived for the changes noted in the viscera and skeleton.

2. The more common findings have been cardiac enlargement, skeletal disturbances involving the long bones, and pneumonia.

3. The roentgen manifestations of changes in bone have been noted to occur



Fig. 15. Radial striations, thickening of the bones, and prominent diploes may be seen in the parietal bones. The frontal bone is also sometimes involved. Only 25.9 per cent of the cases showed any significant change in the skull.

later than the clinical findings of heat, swelling, and tenderness. These destructive bone changes are transitory and may be missed unless serial roentgen examinations are performed.

4. Since sickle-cell anemia is a systemic disease and may manifest itself in many ways, radiologic study should be correspondingly complete.

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SUMARIO

Alteraciones Radiológicas en la Drepanocitemia

Las alteraciones radiográficas observadas en la anemia por células falciformes son clasificadas bajo cuatro encabezados principales: Invasión de las vísceras, incluso el corazón, pulmones, hígado y bazo; alteraciones esqueléticas debidas a hiperplasia de los elementos eritroblásticos; alteraciones esqueléticas debidas a trombosis; y alteraciones esqueléticas debidas a perturbaciones del desarrollo.

En la serie de los AA. la hipertrofia cardíaca fué el hallazgo más frecuente (76.2 por ciento de 42 casos); la hepatización de varios segmentos del pulmón ocurrió frecuentemente, observándose menos a menudo esplenomegalia, hepatomegalia y coleditiasis.

Las alteraciones óseas comprenden engrosamiento de las trabéculas asociado a dilatación de la cavidad medular y adelgazamiento de la corteza. Las trombosis que ocluyen los capilares de los huesos culminan en infarto, necrosis y resorción del hueso.

Los trastornos generalizados del desarrollo se imputan a la desnutrición de los tejidos y las glándulas endocrinas por virtud de la intensa anemia. Las alteraciones comprenden: hábito lineal, emaciación, alargamiento de los miembros, acortamiento del tronco, cifosis dorsosuperior, hundimiento del tórax, prominencia del abdomen, aracnodactilia, y en ocasiones anomalías morfológicas de la cabeza. Estos hallazgos se reflejan en los estudios roentgenológicos.

Se ha observado que las manifestaciones radiológicas de la patología ósea se presentan más tarde que los hallazgos clínicos de ardor, edema e hiperestesia. Dichas alteraciones osteolíticas son pasajeras y pueden pasarse por alto completamente a menos que se ejecuten exámenes seriados con los rayos X.

Dado que la drepanocitemia es una enfermedad orgánica y puede manifestarse de muchos modos, el estudio radiológico debe ser igualmente completo.

Roentgen Therapy for Carcinoma of the Larynx

Fifteen Years Experience¹

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CANCER OF THE larynx is one of the rarer malignant neoplasms encountered in man, comprising 1 to 4 per cent of the entire material. Experience in the treatment of this condition is therefore limited except for a few large cancer centers and several university hospitals renowned for their laryngological work. Time and significantly numerical material are necessary to evaluate any new therapeutic procedure. The era of effective radiation therapy for cancer in this country goes back only to the early thirties and, particularly in the case of the larynx, shortly after Pancoast, Tucker, and others proclaimed the ineffectiveness of irradiation in this disease. They were perfectly right in their assertions before the days of protracted fractionated treatment as evolved by Coutard. In the thirties and early forties, however, there was almost universal acceptance of Coutard's principles and I believe that radiotherapy was given an extensive trial in cancer of the larynx during the decade 1930 to 1940. In more recent years, particularly now, we are again encountering opposition to radiotherapy as an acceptable method of treatment for this condition.

Radiotherapists are in a peculiar position, especially in this country. They play the role of a mendicant, depending on the rest of the profession for their patients. They are often told what to do and how to do it by surgeons and others who have never heard of the inverse-square law, half-value layer, or volume dose distribution, not to mention ion pairs. To make the situation worse, there are many surgeons and specialists who believe that radiotherapy is part of their own specialty and that they should be versed in both,

because only in this way will a patient get an honest, unbiased opinion as to the best type of therapy to be used in a given condition. To debate the question before this audience, which I presume might be a bit partisan, would hardly be cricket, but the problem is pertinent to our discussion. Various reports dealing with the radiological treatment of cancer of the larynx have been discounted because the material was said to have been selected. I ask quite frankly: What radiotherapist selects his material? I believe that in very few instances does he have complete control of the primary material. Patients are usually referred to radiotherapists by surgeons or laryngologists, and real selection of material lies in the hands of the man who controls the disposition of the cases. When this is under the control of one who does both surgery and radiotherapy, the patients may very well be selected with a bias and a leaning *not* in the direction of radiotherapy.

Early in 1931, Dr. Rudolph Kramer, co-author of this paper, otolaryngologist to Mount Sinai Hospital and a surgeon of extensive experience, referred a patient aged fifty-four with an extrinsic cancer of the larynx for radiotherapy. The contraindication for laryngectomy was coronary artery disease. The patient was cured of the cancer but died eight years later of heart disease.

From 1931 until 1942, 108 patients with cancer of the larynx were observed by the authors in the Otolaryngological Service of the Mount Sinai Hospital and in private practice. The sex incidence and age distribution of these cases (Table I) were similar to those reported by other authors. There were 100 males and 8 females, a

¹ From The Mount Sinai Hospital, New York, N. Y. Presented at the Thirty-third Annual Meeting of the Radiological Society of North America, Boston, Mass., Nov. 30-Dec. 5, 1947.

TABLE 1: CANCER OF LARYNX: 108 CASES TREATED AT MOUNT SINAI HOSPITAL, NEW YORK, 1931-42

Age Distribution	
21-30.....	1
31-40.....	7
41-50.....	17
51-60.....	37
61-70.....	31
71-80.....	15
Sex	
Male.....	100
Female.....	8
Primary Treatment	
Surgery.....	28
X-Rays.....	80

ratio of 12.5 to 1. A study of the age distribution showed an increasing incidence from the third decade onwards, with the peak incidence in the sixth and seventh decades.

Every patient was first seen by the laryngologist, who made a detailed study of the extent of the disease. A positive biopsy of squamous-cell carcinoma was also obtained in every case. For reasons discussed in a previous publication, no attempt was made to grade these tumors (1). All patients who were referred for radiotherapy have been included in the present report regardless of the extent of the disease or whether or not treatment was completed.

The classification of carcinoma of the larynx into intrinsic and extrinsic groups on the basis of anatomical location is unsatisfactory. It gives no idea of the extent or severity of the disease; it does not indicate the type of surgery required for its eradication nor the type of response that may be expected from radiation therapy. It gives no idea of prognosis. Quick (2) in 1937, in his Janeway Lecture presented before the Fifth International Congress of Radiology, clearly indicated the shortcomings of this method of classification and suggested a more detailed division of the various anatomical sites of origin for the reporting and analyzing of results from various forms of treatment. Nielsen and Strandberg (3) suggested a method of classifying cancer of the larynx according to stages of the disease, as has been done for cancer of the breast and cancer of the cervix uteri, so that the kind of surgery re-

quired, the type of response to radiation to be expected, and the prognosis might be determined. Their study, however, was limited mainly to cancer arising from the so-called intrinsic larynx. Such a method, if developed further, would probably provide a useful classification.

Because of the lack of a uniform method of classification, we have listed our cases in detail in the latter portion of this paper. However, for statistical purposes, in order to have a basis of comparison with the work of other authors, especially those who have challenged the efficacy of radiotherapy in cancer of the larynx, we have classified our cases as intrinsic and extrinsic according to the criteria described by Hayes Martin (4), even though we realize that this classification is otherwise unsatisfactory. The intrinsic group includes all lesions involving only the ventricles, the true cords, the anterior commissure, the posterior commissure, or the subglottic region. The extrinsic group includes all lesions involving the epiglottis, the aryepiglottic folds, the ventricular bands (false cords), the arytenoids, the piriform sinuses, or the post-cricoid region. We have also classified our material according to the type of surgery that would have been required had it been chosen as the original form of treatment.

Of our 108 cases, 80 were treated primarily by x-rays; 36 were intrinsic lesions and 44 were extrinsic, according to this anatomical classification. The remaining 28 cases were treated primarily by surgery, mainly for didactic reasons and are, therefore, not included in this analysis.

In the various reports on cancer of the larynx, there are marked differences in the relative incidence of intrinsic and extrinsic disease, even allowing for differences of opinion as to what constitutes an intrinsic or an extrinsic lesion. In Jackson's (5) experience, 85 per cent of the carcinomatous growths occurring in the larynx were located on the vocal cords. Martin, on the other hand, found more extrinsic lesions than intrinsic. In our experience, also, extrinsic lesions predominated, but we

were impressed by the fact that the majority of our intrinsic lesions occurred in our private cases, whereas the extrinsic lesions were found to be more common among the clinic patients.

In our five-year analysis of results of treatment by x-ray, 13 cases were indeterminate for the following reasons: 6 died from other causes but with no evidence of cancer; 3 were lost to follow-up but were without evidence of cancer when last seen; 4 failed to return for the completion of x-ray therapy. Of these 13 indeterminate cases, there were 6 in the intrinsic group and 7 in the extrinsic.

TABLE II: CANCER OF THE LARYNX TREATED BY X-RAYS: FIVE-YEAR END-RESULTS, 1931-42

	Intrinsic	Extrinsic
Total cases (80)	36	44
Indeterminate	6	7
Determinate	30	37
Failures	8	22
Well over five years	22	15
Five-year cure rate	22/30 (73%)	15/37 (40%)
Five-year cure rate for all cases	37/67 (55%)	

Sixty-seven cases were suitable for analysis. The results are shown in Table II. Of the 36 intrinsic lesions, 30 were determinate, and in this group there were 8 failures and 22 five-year survivals without evidence of disease. The five-year cure rate was 73 per cent.* Blady and Chamberlain (6), in their article on x-ray treatment of cancer of the larynx, reported a 71 per cent five-year survival rate for their intrinsic cases without metastases and 59 per cent for all of their intrinsic cases including those with lymph node involvement. Martin, on the other hand, reported only a 10 per cent five-year survival rate for his cases treated with x-rays and 50 per cent for those treated by surgery.

Of the 44 extrinsic lesions, 37 were determinate, with 22 failures and 15 five-year survivals. The five-year survival rate was 40 per cent. Blady and Chamberlain reported 25 per cent for their cases treated by x-ray, while Martin reported 11 per cent five-year survival for his irradiated cases and no survivals for those treated by surgery.

TABLE III: CANCER OF LARYNX TREATED BY X-RAYS: FIVE-YEAR END-RESULTS, 1931-42

Total Number, 80; Intrinsic, 36; Extrinsic, 44	
Originally suitable for laryngofissure.....	8
Indeterminate.....	3
Determinate.....	5
Failures.....	0
Well after 5 years.....	5
Five-year cure rate.....	5/5 or 100 per cent
Inoperable or suitable for laryngectomy.....	72
Indeterminate.....	10
Determinate.....	62
Failures.....	32
Well after 5 years.....	30
Five-year cure rate.....	30/62 or 48 per cent

The results just stated are based upon an anatomical classification into intrinsic and extrinsic lesions. If, however, our 80 cases are classified according to surgical indications (Table III), there are 72 cases which were either inoperable or suitable only for total laryngectomy. In 10 of these, results were indeterminate; the five-year survival rate for the remaining 62 was 48 per cent. Eight cases were originally suitable for laryngofissure. These were cases with small, early lesions. Of these, 3 were indeterminate; the five-year survival rate for the 5 determinate cases was 100 per cent. Although this group is too small to be statistically significant, it represents a challenge to Martin's statement that carcinoma of the intrinsic larynx is radioresistant and should be treated only by surgery.

Since our study dates back to 1931, it is possible to report ten-year end-results for the cases observed and treated from 1931 until 1937, inclusive (Table V). There were 38 such cases, of which 33 were determinate; 17 of these patients, or 51 per cent, survived ten years without evidence of disease. Only one of the 17 was originally suitable for laryngofissure. The five-year cure rate for all cases in the 1931 to 1942 period was 55 per cent.

Tracheostomy was performed in 21 cases (Table IV). The indication in every case was laryngeal obstruction. Tracheostomy was not performed for prophylactic reasons in our series. In 11 cases, tracheostomy preceded x-ray therapy; in 9 cases, it was done after the course of radiation therapy was completed. In only one instance was

TABLE IV: CANCER OF LARYNX TREATED BY X-RAY, 1931-42: CASES IN WHICH TRACHEOSTOMY OR LARYNGECTOMY WAS PERFORMED

	Number	Indeterminate	Failures	5-Year Survivals
Tracheostomy	11	1	7	2
Preliminary	1	0	1	0
During treatment	9	0	7	2
After treatment				
Laryngectomy after failure of x-ray therapy	9	1

the course interrupted for tracheostomy. There were only 4 five-year survivals in the cases requiring tracheostomy. In the reports published by Blady and Chamberlain (6) and by Lenz (7, 8), tracheostomy also indicated a bad prognosis. In cases requiring tracheostomy the lesions were, as a rule, advanced. Occasionally, post-irradiation edema of the aryepiglottic folds required tracheostomy, especially if there was a superimposed upper respiratory infection.

TABLE V: CANCER OF LARYNX TREATED BY X-RAY: TEN-YEAR END-RESULTS, 1931-1937

Total Number	38
Indeterminate	5
Determinate	33
10-year survival	17*
10-year cure rate	17/33 or 51%

* Only one suitable for laryngofissure

Total laryngectomy was performed in nine x-ray failures. According to the surgeons performing these operations, no special difficulty was encountered. Only one case, however, was cured by this procedure.

The irradiation technic employed is indicated in Table VI. We have followed the general principles of Coutard (9) of protraction and fractionation in all of our patients with cancer of the larynx since 1931. Our only change in technic began in 1938 when we used higher milliamperage, resulting in a faster rate of reflux per minute, and we started using smaller portals of entry. The course of therapy was usually begun with small doses of 75 to 100 r to the anterior field for the first five days. This was followed by treatments to the right and left lateral fields, with a daily dose of 250 to 300 r measured in air—125 or 150 r to each lateral field. The lateral fields were opposing, without angulation. The cen-

TABLE VI: CANCER OF LARYNX TREATED BY X-RAY: TECHNICAL FACTORS

200 kv.	
Thoraeus filter	
Half-value layer: 1.9 mm. Cu	
Focal skin distance: 60 cm.	
Field size: 5 cm. circle	
or 7 cm. circle	
or 6 × 8 cm.	
12 r per minute	
Over-all treatment time: 30 to 40 days	
Dose	
Anterior neck:	500 to 1500 r (in air)
Right and left lateral neck:	3000 to 4500 r (in air)
Dose at center of larynx:	4000 to 6000 r

tral ray of the x-ray beam was directed through the center of the lesion and appropriate measures were taken to insure that the entire lesion was well within the beam, with an adequate margin of normal, uninvolved tissue. The patients were set up and observed personally by the radiotherapist. They were checked at frequent intervals by the laryngologist. In the majority of cases a membrane reaction of the larynx and hypopharynx developed during the third week of treatment. Radiodermatitis, in the form of a blistering reaction, appeared toward the end of the course of therapy and reached its height a few days later. Complete healing of both radioepithelitis and radiodermatitis occurred in about ten days. With the healing of the membrane reaction, the tumor disappeared in the favorable case. As a rule, the course of therapy was well tolerated, although many patients suffered from pain, cough, hoarseness, and dysphagia during the height of the radiation reaction. From measurements in a water phantom, it has been calculated that the majority of our patients received a tumor dose of 4,000 to 6,000 r. In the favorable case, all symptoms subsided and the larynx presented a normal or an almost normal appearance.

All the determinate cases were further

TABLE VII: CANCER OF LARYNX, 1931-42. INTRINSIC CASES WITH FIVE-YEAR SURVIVAL (73% OF TOTAL)

Location	No. of Cases	Palpable Lymph Nodes	Fixed Cord*
Total	22	1	3
Vocal cord, anterior two-thirds			
Alone	1†
With subglottic region	1†
With subglottic region and anterior commissure	2†
With subglottic region and ventricle	1†
With ventricle	1	..	1
Vocal cord, posterior third			
With subglottic region	1
With posterior commissure	2	..	1
Vocal cord, entire length			
Alone	8
With posterior commissure and ventricle	1
Both vocal cords			
Anterior two-thirds with ventricle	1
One, posterior third; other, middle third	1
Entire length with subglottic region	2	1	1

* Two remained fixed, one movable after therapy.

† Cases originally suitable for laryngofissure.

analyzed, as shown in Tables VII to XI, for details of location and extent of disease. Martin claimed that the site of origin should be ascertained in every case, but we found that this was not possible in the extensive lesions. Coutard believed that the last portion of a neoplasm to disappear during the course of x-ray therapy was the site of origin, but this hypothesis was never proved. The analysis of location and extent was made to determine what factors favor curability or failure. The small number of cases in each group did not clarify this problem.

Prognosis depends upon extent of disease, its location, and the degree of infiltration. From our own analysis, the impression was obtained that the more extensive the disease, the poorer were the results. This was especially true for lesions which involved several anatomical structures. The best results, as might be expected, were obtained with the smaller lesions, and these were usually found on the true cord.

TABLE VIII: CANCER OF LARYNX, 1931-42. INTRINSIC CASES: FAILURES

Location	No. of Cases	Palpable Lymph Nodes	Fixed Cord
Total	8	3	3
Vocal cord, anterior two-thirds with ventricle	1
Vocal cord, entire length			
Alone	1	1	1
With ventricle	1	..	1
With anterior commissure and subglottic region	1*
With ventricle, pre-laryngeal invasion	1	1	1
Both vocal cords, entire length			
With anterior commissure	1
With posterior commissure	1
With subglottic region	1	1	..

* Cured by laryngectomy.

From the point of view of location, the poorest results were obtained with lesions in the post-cricoid region and in the piriform sinus. Lesions of the epiglottis, on the other hand, were favorable, even if the cartilage was destroyed. Involvement of the arytenoid, according to Lenz, indicates a bad prognosis, and he attributed many of his failures to involvement of this structure. On the other hand, Martin singled out the arytenoid as the most favorable location for extrinsic lesions and claimed a more favorable prognosis when that structure only was involved. In our own series, lesions which appeared to originate in the arytenoid were usually extensive when first seen, and of unfavorable prognosis. The prognosis for lesions on the aryepiglottic folds was usually unpredictable and probably depended upon whether the disease extended also to the epiglottis, the piriform sinus, or the arytenoid. The false cord lesions appeared to have a better prognosis. Prognosis for the intrinsic lesions was, in general, much more favorable than for the extrinsic tumors. The best prognosis was obtained with true cord lesions, and, if the disease is limited to the anterior two-thirds of the true cord, cure rates of over 85 per cent should be obtained with either x-ray therapy or laryngofissure. Lesions originating in the ventricle or sub-

TABLE IX: CANCER OF LARYNX, 1931-42. EXTRINSIC CASES WITH FIVE-YEAR SURVIVAL (40% OF TOTAL)

Location	No. of Cases	Palpable Lymph Nodes	Fixed Cord
Total	15	6	4
Epiglottis			
Alone	3
With aryepiglottic fold	2
With anterior commissure and vocal cord	1	1	..
False cord			
With ventricle	2	1	..
With epiglottis and ventricle	2	1	..
Arytenoid			
With aryepiglottic fold and epiglottis	1	..	1
With piriform sinus	1	..	1
Piriform sinus	1	1	..
Extensive			
Half of larynx with fixation of involved side	2	2	2

glottic region may be more extensive when first recognized than lesions originating on the true cords. However, they also respond well to x-ray therapy. Lenz, on the other hand, was of the opinion that subglottic lesions were unfavorable for x-ray therapy because of the tendency of disease in this location to spread down the trachea.

The degree of infiltration is best estimated by the amount of fixation of the neoplasm. Fixation of one cord or of one side of the larynx may be caused by inflammation or mechanically by a large bulky tumor, or it may indicate infiltration of muscles or of the arytenoid joint by cancer. In all of our cases originating in the arytenoid, the cord was fixed. On a few occasions, it was observed that fixation of the cord disappeared during the course of x-ray therapy, from which it might be inferred that fixation was caused by an inflammatory process rather than neoplastic infiltration. Fixed cords were found in 7 of our cured cases and in 16 of the failures. In 5 of the cured cases, the movement of the cord returned after the course of x-ray therapy was completed. In the other 2, fixation of the cord persisted although there was no evidence of disease.

Involvement of cartilage, either by neoplasm or by inflammation, may be a contraindication to x-ray therapy because of

TABLE X: CANCER OF LARYNX, 1931-42. EXTRINSIC CASES: FAILURES

Location	No. of Cases	Palpable Lymph Nodes	Fixed Cord
Total	22	13	13
Epiglottis			
With false cord	1	1	..
With false cord and aryepiglottic fold	1
With false cord, ventricle and vocal cord	1
False cord			
With vocal cord	1
With anterior commissure and opposite false cord	1
Aryepiglottic fold			
With arytenoid, epiglottis, and piriform sinus	1	1	..
Arytenoid			
With ventricle	1	..	1
With false cord, ventricle, vocal cord, and subglottic region	2	1	2
With piriform sinus, vocal cord, and subglottic region	1	1	1
With both vocal cords	1	1	1
Piriform sinus			
Alone	1	1	1
With arytenoid and epiglottis	1	1	..
With arytenoid	1	1	1
Extensive			
Half of larynx	5	3	5
Entire larynx	1	1	1
Post-cricoid	2	1	..

the danger of radionecrosis. The morbidity resulting from radionecrosis of the cartilage is severe. If the patient survives, the cartilage is extruded in the form of sequestra and the end-result is equivalent to a total laryngectomy. Total laryngectomy is, therefore, preferable if the cartilage is obviously involved. An exception to this rule is involvement of the cartilage of the epiglottis, which appears to offer no additional risk to x-ray therapy.

Cervical lymph node enlargement occurred in 23 of our cases, 7 of which were cured cases. In most instances, however, the diagnosis of metastatic disease of the lymph nodes was not confirmed by microscopic examination. In one intrinsic case, enlarged cervical lymph nodes appeared three years after healing of the primary lesion. These nodes responded well to a course of x-ray therapy. The patient

TABLE XI: CANCER OF LARYNX: SUMMARY OF FIVE-YEAR RESULTS WITH PROBABLE SITES OF ORIGIN*

Site	No. of Cases	Cures	Failures
Extrinsic			
Epiglottis	9	6 (67%)	3 (33%)
False cord	6	4 (67%)	2 (33%)
Arytenoid	7	2 (28%)	5 (72%)
Piriform sinus	4	1 (25%)	3 (75%)
Extensive	8	2 (25%)	6 (75%)
Post-cricoid	2	0	2 (100%)
Aryepiglottic fold	1	0	1 (100%)
TOTAL	37	15 (40%)	22 (60%)
Intrinsic			
Vocal cord—			
Anterior two-thirds	7	6 (86%)	1 (14%)
Posterior one-third	3	3 (100%)	0
Entire length	13	9 (69%)	4 (31%)
Both vocal cords	7	4 (57%)	3 (43%)
TOTAL	30	22 (73%)	8 (27%)

* Extension to and involvement of other parts is not indicated.

lived another nine years, when a new carcinoma appeared in the post-cricoid region, from which he died. In another case, a supraclavicular node appeared five years after the primary lesion, which involved the false cord and ventricle, had completely healed. This node responded well to x-ray therapy alone, and the patient is now alive and well fourteen years after the primary lesion was destroyed. In a third case, the primary lesion was located in the epiglottis, false cord, and ventricle, and numerous nodes were present at the time of the original treatment. All of these nodes, except a small group at the lower anterior angle of the neck, responded well to the original course of x-ray therapy. An additional course failed to control this group of nodes which was, therefore, excised surgically. Pathological examination of the excised nodes revealed squamous-cell carcinoma. The patient is now alive and well, more than ten years since the original treatment. In 4 other cured cases, nodes were palpated at the original examination and disappeared after the course of therapy was completed. There was no histologic examination of these nodes to determine whether or not they were involved by carcinoma. Lenz reported cures of epiglottic lesions with lymph node involvement. Nodes were present in 16 of our failures, 3

intrinsic and 13 extrinsic, at the time of the original examination. Here again, very few were examined histologically, but their development and spread during the unfavorable course of the disease in the majority of cases left little doubt about their metastatic nature. The impression gained from this analysis is that the presence of involved lymph nodes indicates a more unfavorable prognosis because such involvement usually occurs late in carcinoma of the larynx and is therefore associated with an advanced primary lesion.

In view of the small number of patients available for study and treatment, parallel series treated by surgery and by x-ray were not carried out at our hospital. A policy of employing roentgen therapy as the primary method of treatment was adopted by the Laryngological Service and Radiotherapy Department at Mount Sinai Hospital. Our results, which have been presented in this article, have demonstrated the wisdom of this policy. There appear to be no statistical data at present which show that surgery has more to offer the patient as regards both recovery and function. We shall, therefore, continue to employ roentgen therapy as the treatment of choice for all cases of carcinoma of the larynx with the exceptions discussed in this article. Our policy is subject to change, however, should proof become available that the patient is better treated by another approach. In the meantime, the results from radiotherapy may be further improved by new technics and other modalities which are being developed and studied.

SUMMARY

1. From 1931 until 1942, inclusive, 80 cases of squamous-cell carcinoma of the larynx were treated with x-rays. Of these, 67 were suitable for statistical analysis.

2. The five-year survival rate for all cases was 55 per cent. On the basis of an anatomical classification, the five-year survival rate for intrinsic lesions was 73 per cent and for extrinsic lesions 40 per cent. On the basis of a surgical classification, the

five-year survival rate was 48 per cent for cases originally either inoperable or suitable only for total laryngectomy.

3. The ten-year survival rate in 33 cases treated from 1931 until 1937, inclusive, for which determinate results were available, was 51 per cent.

4. Tracheostomy performed for laryngeal obstruction, either before, during, or after treatment, indicated a bad prognosis.

5. Total laryngectomy performed in 9 x-ray failures was successful in only one case.

6. The technic, dose estimations, and radiation reactions are outlined.

7. Prognosis depended upon extent and location of disease. The extrinsic lesions were usually much more extensive than the intrinsic when first seen and had a less favorable prognosis.

8. Small early lesions, most of which were located on the true cord, had the best prognosis. Prognosis was favorable also in lesions located on the epiglottis, false cord, or ventricle. An unfavorable outcome may be expected with lesions of the piriform sinus, arytenoid, or post-cricoid region.

9. Involvement of the subglottic region did not, by itself, indicate an unfavorable prognosis.

10. Fixation of the cord occurred in 7 cures and 16 failures. In 5 of the cured cases, mobility of the cord returned after treatment.

11. Involvement of cartilage, either by infection or neoplasm, with the exception of the epiglottis, is probably a contraindication to x-ray therapy because of possible radionecrosis.

12. Cervical lymph node metastasis occurred, as a rule, late in the disease and was usually associated with an extensive lesion. When occurring with curable primary lesions, particularly those on the epiglottis, false cord, or ventricle, cervical lymph node metastases may be successfully controlled by surgical or radiological methods.

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SUMARIO

La Roentgenoterapia en el Carcinoma de la Laringe. Quince Años de Experiencia

De 1931 a 1942, inclusive, tratáronse con los rayos X 80 casos de carcinoma escamocelular de la laringe. De ellos, 67 se prestan para análisis estadístico. La tasa de sobrevivencias de cinco años en éstos representó 55 por ciento. A base de clasificación anatómica, las sobrevivencias de cinco años en 30 lesiones intrínsecas representaron 73 por ciento y en 37 extrínsecas 40 por ciento. A base de clasificación quirúrgica, las sobrevivencias de

cinco años representaron 48 por ciento en los primitivamente ya inoperables o sólo apropiados para la laringectomía total. En 33 casos tratados de 1931 a 1937 inclusive, para los cuales hay datos asequibles, el índice de sobrevivencias de diez años fué de 51 por ciento.

La traqueotomía ejecutada por obstrucción laringea, ya antes o después del tratamiento, o durante el mismo, indicó un pronóstico malo. La laringec-

tomía total, ejecutada en 9 casos de fracasos de los rayos X, sólo obtuvo éxito en uno.

La técnica de la irradiación se basó en los principios de la protracción y la fraccionación, preconizados por Coutard. Calcúlase que la mayoría de los enfermos recibieron una dosis tumor de 4,000 a 6,000 r. En la mayoría de los casos se presentó una reacción membranosa de la laringe e hipofaringe durante la tercera semana de tratamiento. Hacia el final de la serie de terapéutica apareció radiodermatitis en forma de una reacción vesicante. En unos diez días curaron completamente tanto la radioepitelitis como la radiodermatitis. Una vez sanada la reacción membranosa, el tumor desapareció en los casos favorables.

El pronóstico se conformó a la extensión y localización de la enfermedad. Las lesiones extrínsecas eran por lo general mucho más extensas que las extrínsecas al ser observadas por primera vez y tenían un pronóstico menos favorable. El mejor pronóstico correspondió a las pequeñas lesiones tempranas, la mayor parte de las

cuales se hallaban situadas en la verdadera cuerda. El pronóstico fué también favorable en las lesiones localizadas en la epiglotis, las cuerdas falsas o los ventrículos. Cabe esperar un resultado adverso en las lesiones del seno piriforme, el aritenoides o la región postcricoidea. La invasión de la región subglótica no indicó, de por sí, un pronóstico desfavorable.

Hubo fijación de la cuerda en 7 casos de curación y 16 fracasos, reapareciendo la movilidad en 5 de los casos curados, después del tratamiento.

La invasión de cartílagos, ya por infección o neoplasia, con la excepción de la epiglotis, constituye probablemente una contraindicación de la roentgenoterapia, debido a la posibilidad de radionecrosis.

Las metástasis en los ganglios cervicales se presentaron, por regla general, tardíamente en la enfermedad y se asociaron habitualmente con una lesión extensa. Cuando se trata de lesiones primarias curables, en particular de la epiglotis, cuerdas falsas o ventrículos, pueden atacarse con éxito esas metástasis por medio de la cirugía o la radioterapia.

DISCUSSION

(Papers by del Regato¹; Cutler¹; Harris, Kramer, and Silverstone)

W. Edward Chamberlain, M.D. (Philadelphia, Penna.): Dr. del Regato convinced me that he is a very sound therapist who individualizes, who avoids the mistake of treating cancer by rote. He also gave us welcome evidence that, in his Clinic, radiation therapy and surgery are partners and not competitors.

I also like Dr. Cutler's approach to this problem. He spoke of the fact that the radiologist has had to work with the discards. I think that has been all right. I have had the satisfaction for a great many years of realizing that not one single case that came to us for radiation was a case that the surgeons felt they should have had, and I have had some good results in spite of the fact that I was treating discards.

I think we radiologists ought to bear in mind the fact that curability in the case of radiation therapy rests partly on a different basis from curability for the surgeon. For the surgeon, curability is determined, to a large degree, by the ana-

tomic extension of the disease. In other words, if a cancer has not yet involved anatomically certain structures, it can be removed completely and a cure can be obtained, whereas if it has spread to a certain extent it becomes inoperable. All of the statistics that have been presented to you today, and many other good figures, show that the extent and size of the lesion also affect the cure rate for radiology, but not to the same degree because we depend, in part at least, for the ability to get cures, upon the nature of the tumor cell. Therefore we are sometimes successful in cases that the surgeon could not possibly cure. So there is some basis for the radiologist remaining, where he probably will remain whether he likes it or not, in a position of being handed cases by the surgeon.

I cannot wholly agree with Dr. Harris. He doesn't individualize enough, and he has to some extent become a protagonist of radiation therapy. He is a sound therapist, but I don't want to see him continue to be as much of a protagonist of one form of therapy as he appeared to be.

¹ Published in *RADIOLOGY*, October 1948.

Ralph M. Caulk, M.D. (Washington, D. C.):

The essayists are to be congratulated on presenting to us such a large number of cases on which we should be able to draw some conclusions as to treatment of cancer of the larynx.

My own experience at the Warwick Clinic, under Dr. Merritt, parallels largely that of Dr. Cutler. Since 1938, we have followed much the same technic as he has outlined. I hope the essayists won't be offended if I present a brief analysis of all the cases treated in the department of radiation therapy during the years 1933-43, inclusive.

There were 34 consecutive cases; all have been traced, and nearly all of the present survivors examined within the year. Of 19 patients with lesions originating on the true cord, 10 have lived from five to thirteen years and one is living four years. Of this group, 3 cases were too far advanced for a total laryngectomy, and none survived; in 7 patients a total laryngectomy would have been required, and of these, 3 survived; 9 cases could probably have been treated by laryngofissure in the more conservative sense, and in this group there were 8 cures. The failure in the latter group was treated by a total laryngectomy and is a practising dentist in Washington today.

There were 12 cases originating in the laryngeal vestibule, with 5 five-year survivals. These 5 survivals were in a group of 7, in which the disease did not appear to have extended beyond the confines of the anatomical larynx. The remaining 5 vestibular lesions were all advanced, with no survivals.

Three cases originated in the subglottis and all the patients are well, thirteen, nine, and six years, respectively.

I apologize to the essayists for having presented this unorthodox discussion. It is a little added evidence to demonstrate what can be accomplished with roentgen therapy in the treatment of laryngeal cancer.

Maurice Lenz, M.D. (New York, N. Y.): If results of radiotherapeutic or surgical treatment of cancer of the larynx in various clinics are compared with each other, it is presupposed that the extent and location of the cancer in these group are similar. A series consisting predominantly of cases in which the disease is advanced cannot be compared with one in which most of the cases are early. The results of roentgen therapy of Dr. Harris and Dr. Cutler are much better than those of Dr. Martin,² yet they claim that they have,

² Dr. Hayes Martin, in a paper not submitted for publication, gave the cure rate for cancer of the extrinsic larynx by irradiation at Memorial Hospital (New York), over a five-year period, as 10 per cent, and concluded that surgery is the method of choice in all operable cases, intrinsic or extrinsic.

like Dr. Martin, included all cases coming to their respective hospitals and thus present clinical material which is comparable to that of the Memorial Hospital. If that is correct, this superiority can be explained only on a difference in technic, and this is a most difficult factor to evaluate. Results of individualized, unhurried, careful roentgen therapy differ from those of routine treatment administered by inexperienced, over-worked interns. No one would think of comparing laryngeal surgery done by an accomplished attending surgeon with that of a young intern; yet this is unfortunately often done in evaluating radiotherapy. At Presbyterian Hospital, New York, Dr. John Kernan, former Chief of the Laryngologic Service, and I have collaborated in the follow-up clinic for seventeen years. On the basis of this experience we now both agree that roentgen therapy gives as good results, and a better voice, than laryngofissure, in cordal cancer limited to the middle of the vocal cord. In epiglottic cancer treated only by roentgen therapy we obtained five-year to fourteen-year freedom from clinical evidence of cancer in 9 of 28 patients. These results were published in the *Journal of the American Medical Association*, May 10, 1947. We believe that the best results in the treatment of cancer of the larynx will be obtained by a close collaboration of an unbiased laryngological surgeon and a well trained unbiased radiotherapist. I am happy to say that a similar cordial and constructive relationship is now being developed in the Manhattan Eye, Ear and Throat Hospital, New York.

Walter L. Stilson, M.D. (Los Angeles, Calif.):

I want to ask Dr. del Regato a question. In treating carcinomas of the lip with high-voltage radiations is there any attempt made to protect those structures back of the lip?

Dr. del Regato (closing): In the hurried presentation of my summary, I forgot to mention that we do protect the interior of the oral cavity with a heavy shield of lead placed immediately behind the lips and in front of the gingivae.

I want to thank Dr. Chamberlain for his kind remarks. We have been indeed fortunate in collaborating with others. We find it not only profitable, but enjoyable to co-operate with surgeons, including Memorial Hospital trainees, after they have dropped the fatuous pretension of practising radiotherapy "on the side."

We believe that a man is a better cancer surgeon who has an insight into the practice of radiotherapy; we also believe that a man is a better radiotherapist who knows the proper indications and limitations of surgery, and we believe also that it is of advantage to the tumor pathologist to come out of his seclusion and look into the more dynamic and less macabre aspects of cancer. But this

does not mean that anyone may feel qualified to practice these three different and vast branches of endeavor, even though, to facilitate his ambition, he would have the human body arbitrarily divided into more or less unattractive morsels. One cannot direct, one cannot even begin to understand, much less could one practise radiotherapy from the operating room.

Dr. Cutler (*closing*): As I see it, one serious problem is our difficulty in interesting young men in radiotherapy in this surgical age. The tedious placement of patients under x-ray machines twice a day for weeks and the long waiting of months for the result simply are not as exciting as a brilliantly executed laryngectomy. The slow radiotherapeutic method cannot compare with the rapidity and finality of the surgical operation, and the dingy radiotherapy room can hardly compete with the brilliance of the surgical amphitheater. In spite of this, I venture to say that radiotherapy will continue to progress in the hands of those who are temperamentally fitted for it and who have a profound interest in its future possibilities, and

the deep satisfaction of accomplishing cures without mutilation will be more than ample reward for the extra effort.

Dr. Harris (*closing*): The results in radiologically treated cancer of the larynx at Memorial Hospital are at great variance from those reported in the three other papers of this symposium. One must conclude that either the material reported is not comparable or that the methods of treatment account for the great discrepancy in the results. This can be clarified only by a different method of reporting results, where a more detailed classification will be employed and the method of selection of material clearly stated. In my opinion, cancer of the intrinsic larynx can be treated just as successfully by roentgen therapy in patients with lesions suitable for cordectomy or hemilaryngectomy. For more extensive lesions requiring laryngectomy, roentgen therapy appears to give better results both as regards survival and function. This is borne out by the various reports at this meeting and by reports in the literature.



A Study of the Hands of Radiologists¹

NANCY K. BRAASCH and MARGARET J. NICKSON, M.D.

Chicago, Ill.

DURING THE ANNUAL meeting of the Radiological Society of North America in 1946 a study of the hands of volunteering radiologists was undertaken. Brief histories were obtained of each subject, and wax impressions of the finger tips and photographs of the nail-fold areas were made. The purpose of the study was to ascertain the nature and the extent of changes occurring, first, in the epidermal ridges of the fingers, and second, in the minute vessels of the papillae of the corium of the nail fold. It was hoped that some correlation could be made between changes in the two areas studied and the amount of exposure as estimated by the number of years the radiologist has been in practice. It was further hoped that the recognition of such changes would be of benefit in detecting radiation damage before progressive chronic radiodermatitis became obvious.

The first method is one developed and used by Roger Harvey in 1944 (10). It consists of making wax impressions of the epidermal ridges of the terminal segments of the fingers, then studying these impressions for evidences of atrophy, hypertrophy, or other deviations from normal. Harvey found that individuals whose hands had been professionally exposed to radiations often had recognizable changes in the epidermal ridges and that the incidence of change increased directly with years of exposure to radiations (Fig. 1). The wax used in this study was Kerr dental compound, which is brittle at room temperature but softens quickly when warmed in a water bath or under an infra-red lamp. When the warmed surface is soft enough to be indented by a pencil, the finger, which has been dipped into water to prevent sticking, is firmly pressed into the

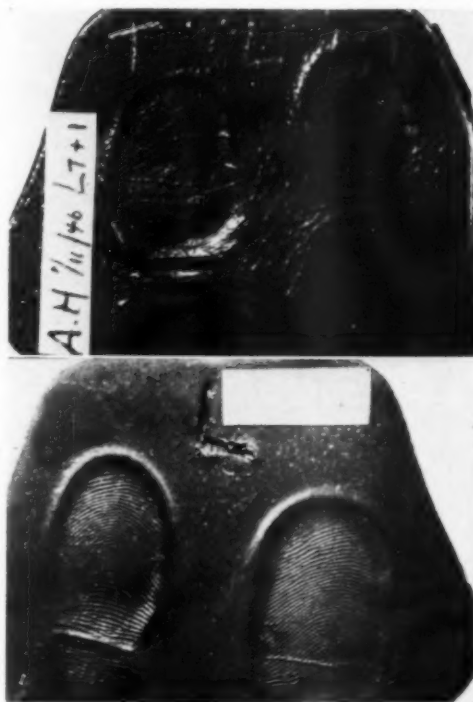


Fig. 1. Impressions in wax of the epidermal ridges of a radiologist (upper) and an unexposed person. Note the generalized atrophy and loss of detail after twenty-seven years of exposure.

compound. After the wax has hardened, it can be read, photographed, or stored as it is. All of the wax impressions collected during this survey were read by Dr. Harvey (see Table I).

The abnormalities found in the impressions of the radiologists in this study varied from areas suspicious for early flattening to areas showing generalized atrophy or proliferative changes. The incidence of abnormality increased steadily with years of experience and was greater than 50 per cent among all who had prac-

¹ Presented at the Thirty-third Annual Meeting of the Radiological Society of North America, Boston, Mass. Nov. 30-Dec. 5, 1947.

TABLE I: PERCENTAGE OF EPIDERMAL RIDGE ABNORMALITIES FOUND BY WAX IMPRESSION METHOD

	Number of Subjects	Percentage with Abnormalities
Harvey's Studies		
Radiologists	1033	68
Controls	1850	1.8
Present Study		
Radiologists	323	48
Argonne laboratory personnel	1880	16
Aged	43	9

tised more than fifteen years and 100 per cent after thirty-five years. The severity of the damage shows a marked increase after fifteen years. The correlation between ridge damage and years of exposure can be seen in Table II.

TABLE II: YEARS OF EXPOSURE TO RADIATION AND FINGER RIDGE CHANGES

Years of Exposure	Number of Individuals	Percentage with Abnormalities
0-5	56	23
6-10	85	28
11-15	48	42
16-20	52	65
21-25	34	74
26-30	26	85
31-35	13	92
36-40	4	100
41-45	2	100

The type of practice appears to have less effect on ridge damage than the number of years of exposure (Table III). Most of the individuals studied are engaged in both therapeutic and diagnostic radiology and roughly half of these show ridge damage. Among those whose work is completely diagnostic 39 per cent show changes.

The type of exposure, whether to x-rays, radium, or both, and its relation to ridge abnormality is seen in Table IV. The subjects who work with x-rays alone and those using both x-rays and radium have

approximately the same incidence of abnormality. Of the 5 men working with radium exclusively, 4 were found to have altered ridges. The fifth, a young man with two years experience preparing radium and radon samples, has normal ridges thus far.

TABLE III: TYPE OF PRACTICE AND FINGER RIDGE CHANGES

Type of Practice	Number of Individuals	Percentage With Abnormalities
Diagnostic and therapeutic	202	52
Diagnostic only	33	39
Therapeutic only	8	50

Questioning as to the use of gloves revealed that few radiologists are without leaded gloves but that their use as a routine practice is exceptionally rare. Most of the radiologists indicated on the questionnaire that gloves were worn "occasionally," "for prolonged exposure," "on right hand only," or, in some instances, after there had been evidence of skin damage. The figures in Table V suggest that hand protection, as it is now practised, is not preventing damage to the epidermal ridges.

The final question on the information sheet was one concerning frequency of radiation surveys. More than half of the subjects who answered this question indicated that surveys with radiation detectors had never been made of their working areas. Some stated that surveys were made routinely but the results were not known to them. Frequency of surveys varied from "every three months" to "every five years." The incidence of ridge damage was 55 per cent among the 86 persons in surveyed areas and 50 per cent among the 112 in unsurveyed areas. Although safety of the working area is not of

TABLE IV: TYPE OF EXPOSURE AND FINGER RIDGE CHANGES

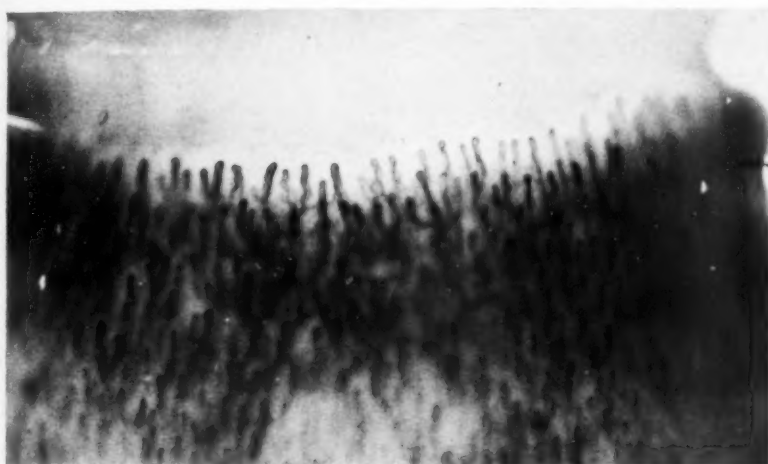
	Number of Individuals	Normal	Localized Atrophy	More than Localized Atrophy	Percentage with Abnormality
X-ray and radium	160	80	57	23	50
X-ray only	140	74	47	19	47
Radium only	5	1	3	1	80

TABLE V: HAND PROTECTION AND FINGER RIDGE CHANGES

Type of Protection	Number of Individuals	Normal	Localized Atrophy	More than Localized Atrophy	Percentage with Abnormalities
Lead gloves	274	142	98	34	48
Leather gloves	4	2	2	0	50
Rubber gloves	2	0	0	2	100
Forceps	2	0	1	1	100
None	12	5	4	3	58

direct concern in this study, it should be remembered that even the best working conditions do not preclude the possibility of excessive hand exposure. More important in this regard is the technic of the individual radiologist.

same vessels can be relocated for repeated observations; the fingers have ample opportunity for incidental exposure during routine radiological procedures; and, finally, because a vast amount of work already has been done on vessels in this area

Fig. 2. Nail-fold area of normal individual. $\times 21.5$

The second method of examination employed in this study is based on the old technic of capillary microscopy (5, 6, 8, 12, 14-16). It has been known for many years that a drop of oil on the skin and proper illumination permit direct observations of superficial vessels of the skin in areas where the epidermis is not too thick. The capillaries, particularly those in the nail fold, have received much attention and their appearance under a variety of conditions of health and disease is well described (1-3, 7, 9-11, 13, 15-17, 21-25).

We have chosen the nail-fold area for our observations for several reasons: the site is readily accessible and can be photographed easily; the precise area and the

under influences other than exposure to radiation. Experimental exposures of other skin areas have indicated that marked capillary alterations can follow relatively small doses (19, 20).

Normally, the nail fold consists of a thin layer of skin extending a short distance over the nail plate in such a fashion as to present a slightly curved area, roughly 2×6 mm., which can be observed under a low-power microscope. The outermost portion is the insensitive epidermis, which appears as a translucent layer of homogeneous tissue. Its free margin makes a wide arc across the field, while its proximal border is a series of scallops formed by the outgrowing papillae of the corium. Usually

each papilla contains one centrally placed vascular loop surrounded by a tissue fluid-filled space. At low magnifications ($\times 20$ to 30), as many as 40 of these may be seen in parallel and are commonly referred to as capillaries of the terminal row (Fig. 2). The afferent or arterial arm has an average diameter of 9 to 12 μ . The efferent or venous limb is slightly wider, measuring from 9 to 20 μ (7). The widening occurs either gradually around the tip or abruptly just proximal to the tip on the arterial side.

vessels, unlike capillaries elsewhere, remain open all of the time. Hence they present a picture which may be as characteristic of an individual as his fingerprints. The shape and pattern of each loop is remarkably constant. Such changes as occur develop gradually over long periods of time and can be followed by serial photographs at intervals of several months or years (Fig. 3).

Photography of this area is not difficult. Special preparation of the finger is not re-



Fig. 3. Photographs of one area of a nail fold. The lower photograph was taken four months after the upper and shows one vessel becoming more tortuous while the others are remarkably unchanged.

When the change of caliber occurs suddenly, there is often what appears to be a small sacculcation in the wall. These sacculcations are thought by us to precede the appearance of dilatation of the venous limb, which in turn seems to precede permanent dilatation or atony of the entire loop. The rate of flow varies from one loop to another. It is generally rapid in the narrow loops and quite slow in the dilated loops. We have always found the direction of flow to be from the narrower to the wider segment, and never have we observed a reversal, as is so common in arteriovenous anastomoses. Nor have we ever seen these vessels open and close, as they seem to do in other sites. Our observations support those of Carrier (4) and of Crawford (5), who found that nail-fold

quired. In the laboratory we ask each subject to scrub gently to remove accumulated debris. Counts made before and after scrubbing have not shown significant differences. The subject then sits with his arm resting at about mid-thoracic level and his finger in a clamp on the stage of the microcamera. A drop of mineral oil or immersion oil is placed on the nail fold, and the vessels are then brought into focus by moving the adjustable stage. The microcamera used in this study is a shop-made funnel-shaped metal box 18 inches long. At the narrow end is a camera shutter and a 24.3-mm. Bausch and Lomb objective. At the upper end is a 4 \times 5-in. ground glass screen on which the image is focused. Under these conditions one obtains a 4 \times 5-in. film on which the capil-

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larities appear at 21.5 times their original size. Sufficient light for photography at 1/10 of a second is produced by mounting a 250-watt projection lamp on each side of the camera. The light is passed through a heat-reducing glass filter (Corning 3966). Eastman Contrast Process Orthochromatic film has been used most satisfactorily. With the same arrangement and exposure time, one can also use Eastman Kodachrome film with good results.

Using this method we have studied nail-fold capillaries of four groups of people:

1. Radiologists
2. Personnel of the Argonne National Laboratory
3. Students and employees of the University of Chicago
4. Aged persons

The first group consists of persons whose opportunities for excessive hand exposure are relatively great. The radiologists studied in 1946 had been in practice for periods ranging from a few months to forty-five years. Many of the older ones had gross evidences of chronic radiodermatitis and some gave histories of acute and severe reactions. The second group consists of persons whose opportunities for exposure are limited. These are the biologists, physicists, chemists, technicians, and office workers of the Argonne National Laboratory. At the time this study was made, few of them had worked longer than three years in the laboratory. Most of them had no history of previous exposure to radiations. The third group consists of young non-exposed individuals. The fourth group consists of old non-exposed individuals. This fourth group was included because of the possibility that age rather than radiations may have induced the changes found in the capillaries and underlying vessels of the older radiologists (18) for it is, of course, true that the subjects with the longest periods of exposure were also the older ones.

The photographs of the nail-fold capillaries were examined by mounting the negative on a light box. The number of

TABLE VI: NAIL-FOLD ABNORMALITIES AMONG RADIOLOGISTS AS COMPARED TO OTHER GROUPS

	Radiologists	Laboratory Personnel	Aged	Students
Number of individuals	152	97	41	165
Number with abnormalities	138	86	40	113
Distribution of Abnormalities	%	%	%	%
Tortuosity	91	87	77	50
Sacculation	28	29	15	16
Dilatation	55	73	57	44
Plexus distortion	9	3	15	2
Branching	12	1	10	5
Papillary defects	12	5	10	6

terminal row vessels occurring in 4 mm. of nail fold was determined. It was found that the count usually fell between 30 and 40 and was not affected by either exposure to radiations or by old age. We then studied the photographs for deviations from the simple normal pattern commonly found in healthy young adults. Six changes appeared repeatedly. These were taken as points on which the films could be judged.

1. Tortuosity of the terminal loop
2. Sacculation of the terminal loop
3. Dilatation of the terminal loop
4. Distortion of the superficial plexus
5. Branching from the terminal loop
6. Defects of the papillae of the corium
 - (a) Atrophy with flattening of the border and loss of vascular components
 - (b) Hypertrophy with the development of macropapillae

The first three changes occurred commonly in all groups studied. Tortuosity has been found so frequently by other authors that it has almost succeeded in being considered a normal finding. Its low incidence in the control group, however, and particularly in young people, tends to support the stand that only straight regular vessels are truly normal.

There appears to be no single abnormality pathognomonic of radiation damage

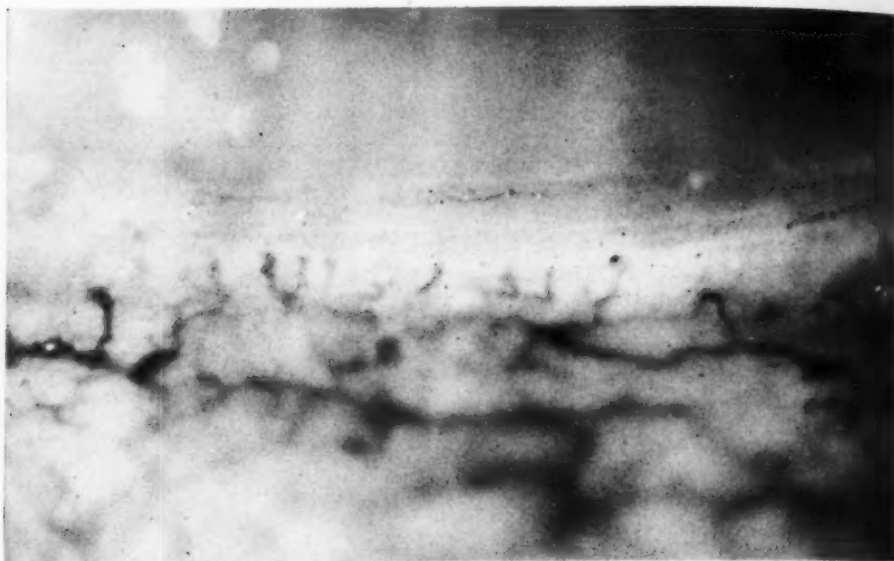


Fig. 4. Nail fold of a radiologist who has practised for forty years. Note the flattened border of the corium and the scarcity of capillary loops. The plexus vessels are irregularly widened and distorted. This subject has clinical evidence of chronic radiodermatitis.

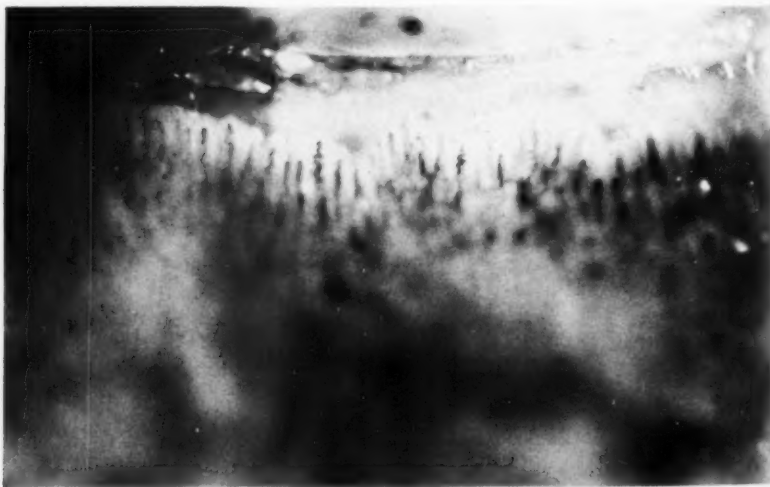


Fig. 5. Two macropapillae in the nail fold of a physicist with a history of many years of exposure to radiation. This subject has had several epitheliomas removed from the skin of the fingers.

(Table VI). Tortuosity occurred in 91 per cent of the radiologists. But it occurred nearly as frequently in the laboratory personnel and in the aged. Sacculation and dilatation seem equally unreliable as indicators of radiation damage. Plexus dis-

tortion, branching, and distortions of the papillae were more common in radiologists and the aged than in the other groups (Figs. 4 and 5). If one considers all abnormalities and calculates the mean number per person, it is apparent that for

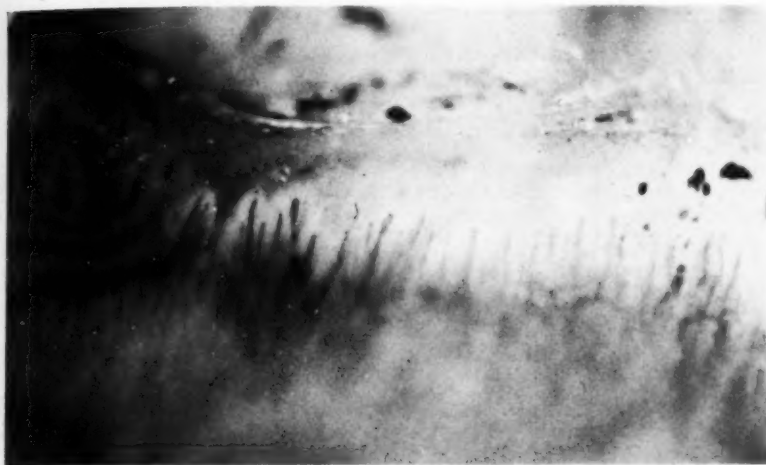


Fig. 6. From a 96-year-old man with no known exposure to radiation. The vessels and papillae are essentially normal.

radiologists this figure advances steadily until it exceeds that of the unexposed aged (Table VII).

TABLE VII: NAIL-FOLD ABNORMALITIES PER INDIVIDUAL IN VARYING AGE GROUPS

Group and Age Range	Mean Number of Abnormalities
Students: 15 to 30	1.22
Aged: 63 to 97	2.07
Radiologists	
21 to 30	1.33
31 to 40	1.92
41 to 50	1.89
51 to 60	1.89
61 to 77	2.94

It is our opinion from these capillary studies that no abnormality of the minute vessels of the nail fold can be considered pathognomonic of radiation damage. The changes which occur are morphologically indistinguishable from the changes found in apparently normal individuals, but the changes appear to be induced at an earlier age among radiologists than among unexposed persons. No radiologist who had practised thirty years or more had normal nail-fold vessels, although normal vessels were seen in aged persons (Fig. 6).

SUMMARY

A study was made of the hands of radiologists by two methods. The first con-

sisted of making wax impressions of the finger ridges and examining these for deviations from normal. The incidence of abnormality among radiologists was 48 per cent as compared to 1.8 per cent of a previously reported unexposed group. The incidence of ridge abnormality increased with years of exposure to 100 per cent for those who had practised more than thirty-five years. The use of lead gloves was not a routine practice in most cases. The incidence of ridge abnormality was 48 per cent among those who professed to use lead gloves.

The second method of study consisted of photographing the nail-fold area and examining these photographs for abnormalities of the terminal vessels, the superficial vascular plexus, and the papillary border of the corium. In general, the same abnormalities were found in a group of aged persons without exposure to radiation as in radiologists, but less frequently than in radiologists after many years of exposure. The most reliable criteria of damage seem to be (1) distortion of the superficial vascular plexus, (2) branching of the vessels of the terminal row, and (3) distortion of the papillary border of the corium. These changes, which may be induced by repeated exposure to radiation, like other biological effects of radiation, cannot be

distinguished from similar findings in very old persons.

ACKNOWLEDGMENT: The following persons assisted greatly in preparing material for this report: J. A. McIntosh, T. Turlentes, and Louise Warner.

The studies of the aged persons were possible because of the co-operation of the directors and members of the Chicago Home for Incurables and the Home for the Jewish Aged.

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SUMARIO

Estudio de las Manos de los Radiólogos

En este estudio de las manos de radiólogos se utilizaron dos técnicas distintas. La primera consistió en hacer impresiones en cera de los surcos digitales, estudiándolas después en cuanto a desviaciones de lo normal. La incidencia de anomalías entre los radiólogos fué de 48 por ciento, comparada con 1.8 por ciento en un grupo previamente comunicado sin exposición a la radiación. La incidencia de anomalías en los surcos acrecentó con los años de exposición, llegando a 100 por

ciento en los que habían ejercido la profesión más de 35 años. El empleo de guantes de plomo no era de rigor en la mayor parte de los casos. La incidencia de anomalías digitales fué de 48 por ciento en los que, según dijeron, usaban guantes de plomo.

La segunda técnica de estudio consistió en fotografiar la zona del repliegue ungüal y en estudiar las fotografías en busca de anomalías de los vasos terminales, el plexo vascular superficial y el borde

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papilar del corion. Se aplicó también esta técnica en otros tres grupos, a saber: el personal del Laboratorio Nacional Ar-gonne (representando personas con limi-tadas ocasiones de exposición a la radia-ción), un grupo de jóvenes sin la menor exposición, y un grupo de personas de edad sin exposición. En general, obser-váronse las mismas anomalías, aunque menos frecuentemente, en el grupo de personas de edad que entre los radiólogos

con muchos años de exposición. Las señales más fidedignas de lesión parecen ser: (1) deformación del plexo vascular superficial; (2) ramificación de los vasos de la hilera terminal, y (3) deformación del borde papilar del corion. Estas alter-aciones, que puede provocar la exposición repetida a la irradiación, al igual que otros efectos biológicos de la misma, no pueden distinguirse de los hallazgos semejantes en personas de mucha edad.



A 35-mm. Unit for Cinefluorography¹

J. S. WATSON, JR., M.D., and SYDNEY WEINBERG

Rochester, N. Y.

DURING THE PLANNING of the 35-mm. apparatus here described, the Department of Radiology was fortunate in having access to a 16-mm. unit assembled some ten years previously by engineers of the Eastman Kodak Co. under the supervision of Mr. Rex Willsey. Basically it consists of an $f/0.81$ cine lens of 4.1 cm. focal length made at the Eastman factory after a lens formula of B. Luboshez,² and a Cine-Kodak Special camera with an unusually rapid pull-down or intermittent, which at normal camera speed moves the film into place in about one-fourth the time required by the conventional film shuttle.

This Kodak experimental unit was designed primarily for ease and simplicity of operation. Recently the lens has been coated, and a Patterson B2 screen installed, increasing the photographic speed by more than 50 per cent. Probably no combination of this sort, using a refractor lens, requires less x-ray energy to expose the film.

The new 35-mm. unit, while slightly inferior in photographic speed, embodies certain well tried technical refinements which it is hoped will give better pictures in some of the more difficult applications of cinefluorography.

In spite of 16-mm. convenience and economy, 35-mm. film would appear to have a number of advantages in this field, not the least being that the larger film can be worked over on an optical printer without appreciable loss of quality. In this way the rate of action on the motion picture screen can be speeded up or slowed down. Many other changes can be made by optical printing, but slowing the action is perhaps the most useful, being nearly

indispensable in the case of scenes which have had to be taken at less than normal camera speeds because of poor illumination.

Optical printing of one sort or another has been in use since the earliest days of cineradiography, when serial radiograms were first rephotographed on cine film in an animation camera. In this kind of transfer each negative image is generally repeated a number of times on the print, even when the subject of the film is a slow-moving viscus (1). The process of positioning or registering unperforated negatives, however, is slow and laborious, and the resulting motion picture is not always as steady as one would wish. In the modern optical printer, on the other hand, where cine film is printed on cine film, the steadiness of the print is limited only by the accuracy of the film perforations. Due to the relative unsteadiness of the smaller film in this respect, 16-mm. to 16-mm. optical printing is not generally considered satisfactory, whereas 35-mm. to 35-mm. and 35-mm. to 16-mm. optical printing are highly perfected and available in most film centers.

If the larger film is decided on, partly for reasons of steadiness, it would be inconsistent to employ any but the most reliable camera movement, preferably with register pins, even though the advantages of the quick pull-down would have to be abandoned. The exposure time lost because of the slower film movement (about 45 per cent at normal camera speeds) is made up by increasing the output of the x-ray machine, while the resulting excess dose to the subject is eliminated without

¹ From the Departments of Radiology and Medicine of the University of Rochester School of Medicine and Dentistry. Aided by a Special Research Fund.

² This pioneer in the field of cineradiography and cinefluorography designed and had made in 1928 a number of fast lenses ranging in speed from $f/0.85$ to $f/0.67$, several years before the appearance of the Zeiss R-Biotar series. Unfortunately the Luboshez lenses were never manufactured in quantity.

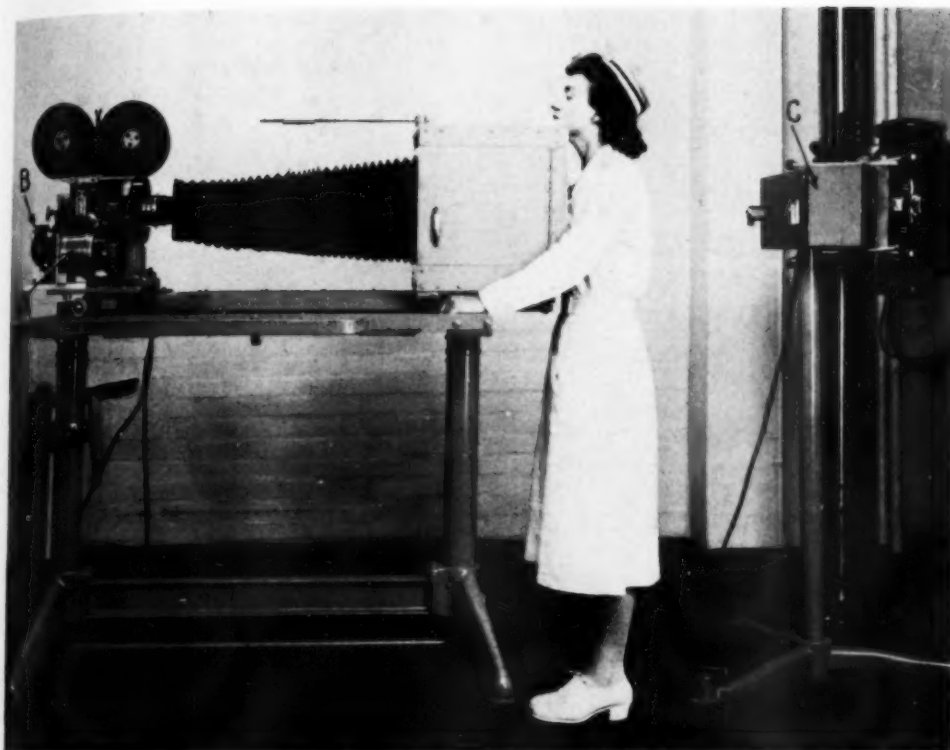


Fig. 1. Thirty-five-millimeter cinefluorographic unit. Camera at left with camera drive motor (B) and repeater motor (A). Lead shutter mechanism (C) between x-ray tube and subject.

loss of useful energy by interrupting the x-ray beam during the pull-down or closed-shutter phase. For this purpose, a lead shutter synchronized with the camera (2) can be set up in front of the x-ray tube. Reynolds (3) recommended interrupting the primary transformer circuit, but the engineering difficulties involved in this method are pretty severe.

It will be noted that in spite of all this maneuvering, roughly twice as much x-ray intensity is needed to expose film in the 35-mm. as in the 16-mm. apparatus. On the other hand, the more critical ratio of patient-dose to film-exposure remains practically unchanged. What would seem to make the extra trouble worth while is the anticipated reclamation of scenes taken at slow camera speeds; for in the present state of cinefluorography these would include nearly all scenes requiring penetra-

tion of parts of the body denser than the thorax.

DESCRIPTION OF 35-MM. APPARATUS

The 35-mm. unit is assembled from available pieces of equipment as follows: Patterson E2 fluoroscopic screen, $14\frac{1}{2} \times 14\frac{1}{2}$ inches; Bell and Howell standard 35-mm. camera; Zeiss R-Biotar lens f. 5.5 cm., $f/0.85$.

The camera is altered to receive the lens by removing the turret front and the forward shutter leaf and by cutting away a segment of the turret housing on the right side. It may be well to mention that the lens does not cover the 35-mm. frame right to the corners, but that the gradual falling off in sharpness and illumination outside the central 15-mm. circle is not considered serious enough in practice to outweigh the advantages of the larger film.

The camera and camera drive motor (Fig. 1) are mounted on the tail stock of a 36-inch lathe bed, so that the camera can be moved near the screen for close-ups or drawn back to cover a larger area. The lathe bed is marked off, and the lens settings are predetermined for each distance. Lead diaphragms rather than cones are used to confine the x-ray beam to the particular screen area desired. The entire assembly, including the screen, is supported on a mobile pedestal, adjustable for height. By using a detachable first surface mirror and changing the position of the screen, subjects can be fluorographed lying down.

The lead shutter for interrupting the x-ray beam (Fig. 2) consists of an aluminum disk similar in design to the rotary camera shutter. It is driven by a 110-volt A.C. repeater motor (Selsyn type), which keeps time with a similar motor geared directly to the camera drive mechanism. A 185-degree segment of the shutter is cut away, while the remaining 175-degree segment is covered with 2 mm. of lead. This ratio takes care of slight variations in synchronization.

The camera and the mate to the repeater motor for the lead shutter are driven by a Lee variable speed 110-volt A.C. motor with an 8:1 reductor head. This permits a camera speed range of 6 to 32 frames per second. A potentiometer is used for gradual starting of the camera and the relatively heavy lead shutter, several seconds being required to attain normal speed. In stopping the assembly, a switch cuts the separate power supply to the repeater motors at the same time that the camera drive motor is shut off, thus allowing the lead shutter to free wheel to a stop independently of the camera. Starting and stopping are done by remote control.

Most of the pictures so far have been made with a General Electric CRT 1-2 tube. Kilovoltages have ranged from 70 to 95 kv.p., with occasional use of 110 kv.p. by permission of the manufacturer. Controls include a ganged deck switch, allow-

ing rapid change-over from 3 ma. for preliminary fluoroscopy to radiographic milliamperages of 25, 50, 100, 200, or 400 ma. Fluoroscopy helps in positioning and timing.

In order to give a general idea of the relative exposure factors involved, which would be independent of variations in output of particular x-ray machines, graded series of single exposures were made of a 22-cm. chest on 16-mm., on 35-mm., and on regular 10 × 12-inch screen films with medium speed screens. A stationary grid was used in all cases. Voltage, amperage, filtration, and target-screen distance were kept constant, and films were developed to approximately the same gamma. Matching negatives were obtained with the following time factors:

10 × 12 screen films.....	1
16-mm.....	1.5
35-mm.....	2

In the case of the 35-mm. negatives, the falling off of density at the edges and in the corners of the image makes matching rather arbitrary. If the central 15-mm. circle only is considered, the two lenses are found to be much more nearly alike in speed.

To translate single exposure factors into motion picture terms, it is necessary to know how long each frame is at rest in the film shuttle. The following table shows how this time interval varies in the two cameras at different camera speeds:

Camera Speed	35-mm. Camera	16-mm. Camera
16	1/34 sec.	1/18 sec.
32	1/68 sec.	1/43 sec.
64	1/136 sec.	1/128 sec.

SUMMARY

1. An experimental 16-mm. apparatus for cinefluorography having an unusually rapid film movement and fast lens is described.

2. Through experience with this apparatus it was found that scenes of parts of the body denser than the thorax must often be taken at slow camera speeds. Such scenes can be improved by optical printing.

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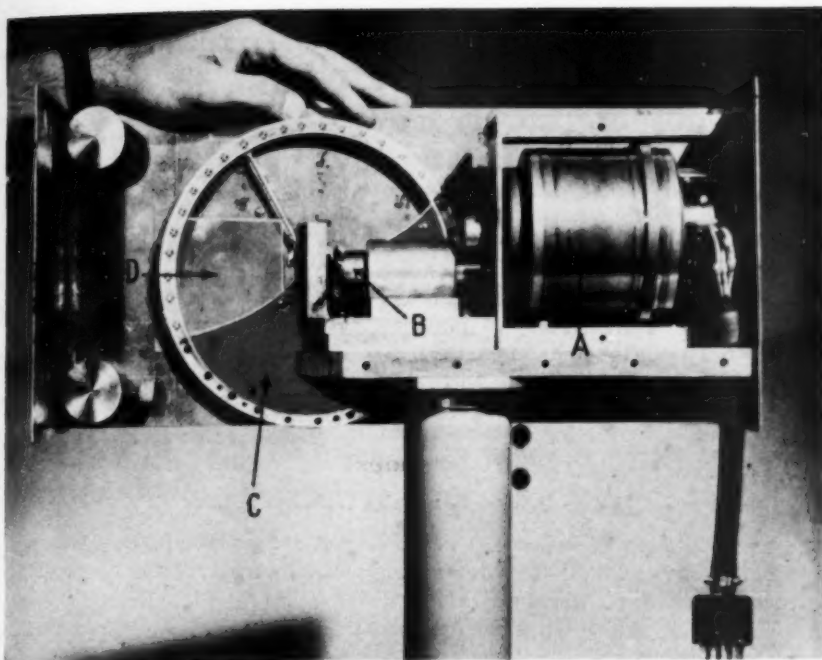


Fig. 2. Lead shutter with safety cover removed. A. Repeater motor. B. Mitre gear drive. C. Leaded segment. D. Plastic window.



Fig. 3. The two camera assemblies: 16-mm. camera with $f/0.81$ lens at left; 35-mm. camera with $f/0.85$ lens at right. A popular $f/1.5$ lens is shown below for comparison.

3. Thirty-five millimeter negatives are more satisfactory than 16-mm. for optical printing, even when the final projection print is to be on 16-mm. film.

4. A new 35-mm. apparatus has the following special features:

- (a) Conventional camera movement with register pins for maximum steadiness.
- (b) Synchronized lead shutter to interrupt the x-ray beam during movement of the film.
- (c) Provision for taking close-ups of screen image comparable to "spot film" views of restricted areas, only magnified.
- (d) Provision for taking scenes of re-

cumbent subjects by the use of a 45-degree mirror.

NOTE: The authors gratefully acknowledge the aid and advice of George H. Ramsey, M.D., J. J. Thompson, M.D., and Frank Dreisinger of the Department of Radiology and of Harry L. Segal, M.D., of the Department of Medicine.

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SUMARIO

Aparato de 35 mm. para la Cinefluorografía

Descríbese un aparato experimental de 16 mm. para cinefluorografía, dotado de un movimiento sumamente rápido para películas de una lente veloz. Practicando con este aparato se observó que hay que tomar a poca velocidad de la cámara las vistas de las partes del cuerpo que son más densas que el tórax. Esas vistas pueden mejorarse por medio de la impresión óptica.

Aun cuando la impresión definitiva de la proyección vaya a ser en 16 mm., las negativas de 35 mm. resultan más satisfactorias que las de 16 mm. para impresión óptica.

El nuevo aparato de 35 mm. posee las siguientes características propias:

- (a) Movimiento convencional de la cámara con clavijas de registro para obtener una estabilidad máxima.
- (b) Obturador sincronizado de plomo para interrumpir el haz de rayos X durante el movimiento de la película.
- (c) Medio de obtener vistas próximas de la imagen en la pantalla, comparables, aunque ampliadas, a las "radiografías instantáneas" de zonas limitadas.
- (d) Medio de obtener vistas de sujetos recostados por medio del empleo de un espejo de 45 grados.

EDITORIAL

The Serious Limitations and Erroneous Indications of Biopsy in the Diagnosis of Tumours of Bone

The essential evidence respecting a bone tumour is whether it is simple or malignant. The clinical history and condition of the patient at the first examination, though often of vital importance in diagnosis, may contribute little to elucidate the problem. Because of this and because the histology of normal tissues is characteristic, while pathology is associated with considerable change in the cellular and intercellular structure, we turned to histology for conclusive evidence. A widespread belief prevails in the infallibility of this evidence. Indeed, so firmly is this belief held that, if the subsequent history of the case differs from the forecast given, it is the pathologist, however eminent, who is blamed for the erroneous interpretation, rather than the vagaries of the histological material. Consequently, certain leading authorities hold that biopsy affords a means of "fully proving" the nature of bone tumours and always resort to biopsy prior to any major surgical measures, notwithstanding Ewing's advice that "few surgeons realise the limitations in the histological diagnosis of bone tumours and the conditions which simulate or accompany them."

An examination of the reports on the histology of bone tumours reveals the indecision which the appearances produce. For, whereas in the descriptions of normal tissues we see the use of definite terms, such as myxomatous, fibrous, cartilaginous, or osseous, in the description of pathological tissue we see the indecisive terms mucoid, fibroid, chondroid, osteoid, muco-fibroid, fibrochondroid, chondrosteoid, etc., terms which permit of considerable latitude of expression by different observers. The

medical student is usually taught his pathology, histology, and radiology from well established lesions and does not realise how closely simple and malignant lesions simulate one another in the early and, indeed, in some cases, in the late stages.

Biopsy involves the risks of anaesthesia, surgical exploration, and all that it means in additional damage, dispersal of tumour cells, destruction of the scaffolding on which repair may be built up, etc., and the mental and physical pain associated with it—risks sometimes resulting in the death of the patient with a simple lesion, which could possibly be regarded as negligible or justifiable if the evidence so obtained were reliable. Consequently, as I have pointed out more fully elsewhere,¹ we ought before biopsy to satisfy ourselves with the answers to several questions: (1) Can the findings of biopsy be relied upon? (2) How should the biopsy be performed—by needling, punch, or real exposure and inspection? (3) Does any form of biopsy permit us to watch sufficiently the evolution of a tumour to establish its nature? (4) Will biopsy enable us to get an early correct diagnosis? (5) If the histology is reported as indicating malignancy, would the surgeon amputate a limb for an early lesion which on clinical and radiographic evidence is uncertain, and, if he would not, what can justify the risks of the surgery of biopsy? (6) Does the evidence of trauma assist in diagnosis or influence biopsy? (7) Has everything else been done to determine the nature of the tumour?

I have ventured in the paper cited above to show that the answers to these questions

¹ Proc. Roy. Soc. Med. 41: 225-236, April 1948.

are opposed to biopsy being a justifiable procedure. No matter which method is used, it is unreliable; has resulted in delay in establishing the correct diagnosis, and in the destruction of limbs with lesions which, left alone, would have resolved; is often, as far as the welfare of the patient is concerned, purposeless and even dangerous; and because of the duration of the interval between onset and biopsy, affords material the initial changes in which cannot be estimated. It is too often resorted to before adequate investigation has been done.

We have learned so much about the radiographic features of bone and joint pathology that, without causing the patient pain, in all but a small minority of cases we can secure an accurate diagnosis in this way, particularly when the findings are judged with the clinical evidence over a period, the duration of which is determined by the rapidity of the changes.

Classification on cellular structure is of but academic interest and unreliable. Spectacular radiographic appearances, unfortunately for the patient, appear to incite surgical exploration for histologic preparations, in spite of the serious risks which have been indicated, the chief of which is erroneous interpretation.

Not the least value of radiology is its use in watching the progress of a lesion during and after the exhibition of chemical and physical agents. The response seen by radiography to follow antisyphilitic medication is a better indication of the nature of the lesion than its histology or than can be obtained by blood reactions. The response of certain tumours to such agents as irradiation, gonadectomy, stilboestrol, Coley's fluid, etc., give us the hope that some specific remedial treatment will be soon found for sarcoma.

This places the responsibility of interpretation where it belongs, *i.e.*, on the radiologist. Too often in the past his reports have been inconclusive, in the hope that a biopsy would be made which would supply evidence agreeing with one of the tentative suggestions made, and have given the clinician little or no guidance in treatment on which he could rely. Because radiologic reports have been so vague, clinicians have sought refuge in biopsy, but this action merely secures other unreliable evidence. Alas! a summation of unreliable reports does not secure accuracy. We have proved that little is to be gained by precipitate amputation. Amputation at the earliest possible moment does not ensure cure, but more careful clinical and radiological investigation of these cases will reduce the number of cases "cured by amputation." Ultimate resolution of a bone tumour which had clinical and radiographic (and even histologic) features which were interpreted as indicating malignancy is the best evidence of benignancy or removal of the destructive influence.

Museum specimens of lesions in amputated limbs do not assist us to a classification which helps us to an early diagnosis, for, in the words of one leading pathologist who was referring to a lesion which had the distinctive radiographic features of an ossifying haematoma and resolved as such, "Had the leg been removed in my case, it would have been impossible to prove that it was not a sarcoma cured by amputation."

We shall have a long way to travel before we arrive at a cure for sarcoma if we continue to regard prompt amputation as the essential treatment.

JAMES F. BRAILSFORD, M.D.
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ANNOUNCEMENTS AND BOOK REVIEWS



Dr. William D. Coolidge

WILLIAM D. COOLIDGE LABORATORY

More than 1,500 persons, including some 200 radiologists en route to the Annual Meeting of the American Roentgen Ray Society, witnessed the dedication, on Sept. 13, of the William D. Coolidge Laboratory of the General Electric Corporation at Milwaukee, Wis. It is especially fitting that this, the world's largest x-ray developmental laboratory, should bear the name of Dr. Coolidge, not alone because of his long association with the General Electric Corporation but more especially for the part he has played in the development of x-ray equipment. Outstanding among his achievements is the Coolidge x-ray tube based on the demonstration of Langmuir, some thirty-five years ago, of the possibility of pure electron discharge in a high vacuum.

This radical departure from previous equipment greatly enlarged the potentialities of radiology in both its medical and industrial applications. It was,

however, but a beginning of the contributions of Dr. Coolidge to the specialty of radiology. Seeking to develop x-ray tubes with higher voltages, he devised a sectional tube, constructed on the "cascade" principle, in which the total voltage is divided between a series of sections. This principle is applied today in the building of tubes for operation at a million volts or more. The first units of this type, developed by Dr. Coolidge and his associate, Dr. Ernest E. Charlton, were for hospital use in cancer therapy and were designed for 800,000 volts. Later units were the 1,000,000-volt and 2,000,000-volt equipments used in both medicine and industry.

No modern hospital today is without x-ray equipment embodying Dr. Coolidge's ideas, and a deep debt of gratitude is owed him not alone by the physician and surgeon, but by an unnumbered host of men and women to whom even his name may be unknown.

Dr. Coolidge's address on the occasion of the dedication of the new laboratory, "Highlights of the Past and a Challenge for the Future," will appear in the December issue of RADIOLOGY.

THE ROLLIN H. STEVENS BIOCHEMISTRY LABORATORY

At the formal dedicatory exercises of the new home of the Detroit Institute of Cancer Research, Oct. 5, 1948, the late Dr. Rollin Howard Stevens, long associated with the Radiological Society of North America and at one time its President, was honored by the dedication of the Biochemistry Laboratory of the Institute in his memory.

The Detroit Institute of Cancer Research, organized in 1941, represents the culmination of a long continued effort on Dr. Stevens' part. His entire experience as a practising radiologist and investigator in ways and means of combating cancer led him to the conviction that the only hope of its conquest lay in research. So strong did this conviction become that a large part of his energies during the later years of his life was devoted to the organization in Detroit of an Institute to be devoted to this purpose.

Dr. Stevens did not live to see the complete fulfillment of his vision—an adequately housed, fully staffed research institute—but those whom he inspired have carried on the work. In the dedication of the Institute as a whole and of the Rollin H. Stevens Biochemistry Laboratory in particular, they paid tribute to a man of vision, indomitable will, and courage.

Speakers at the dedicatory exercises were; Harvey M. Merker, Sc.D., President of the Board of Trustees of the Detroit Institute of Cancer Research, Milton A. Darling, M.D., Chairman of the Cancer Committee of the Wayne County Medical Society, and Douglas Quick, M.D., of Memorial Hospital, New York.

THE THIRD INTER-AMERICAN CONGRESS OF RADIOLOGY

As previously announced in RADIOLOGY, the Third Inter-American Congress of Radiology will assemble in Santiago, Chile, Friday, Nov. 11, to Thursday, Nov. 17, 1949.

Participants in the Congress will be classified as follows: *Honorary members*, those appointed by the Chilean Radiological Society; *Titular members*, those radiologists who apply and are accredited by their respective societies; *Associate members*, physicians who are not specialists in radiology, technicians, experts, manufacturers of electro-medical equipment, and others who may apply, the latter being limited to three for each titular member.

James T. Case, M.D., of Chicago, has been delegated as regional secretary for the United States.

The Committee on International Affairs of the American College of Radiology, consisting of Ross

Golden, M.D., Hugh F. Hare, M.D., Maurice Lenz, M.D., Charles L. Martin, M.D., Frazier J. Payton, M.D., Juan A. del Regato, M.D., M. C. Sosman, M.D., Calvin L. Stewart, M.D., and James T. Case, M.D., has been designated by the American College of Radiology, the American Radium Society, the American Roentgen Ray Society, and the Radiological Society of North America as their official committee on arrangements.

Application for membership in the Congress should be made at the earliest possible date, so that the Chilean Organizing Committee may have an adequate estimate of the extent of the preparations they need to make. The membership fee of twenty dollars should accompany the application.

The official languages will be Spanish, Portuguese, English, and French. We are informed that provision will be made whereby earphones will be available for all who desire them, so that addresses given in Spanish may be heard simultaneously in English, or vice-versa. This means, of course, that those members wishing to offer English contributions to this Congress will either arrange to have them translated into Spanish here or will get them into the hands of the regional secretary to be forwarded to the Congress headquarters in Santiago, Chile, sufficiently in advance of the meeting to permit time for translation.

The official subjects for consideration at the Congress will be the following:

- A. Radiological Exploration of the Cardiovascular System with Opaque Material.
- B. Diagnosis and Simple Radiological Exploration of the Skull, which has been subdivided as follows:
 - (a) Skull in General
 - (b) Sella Turcica
 - (c) Orbits
 - (d) Paranasal Cavities
 - (e) Temporal Bone
- C. Radiation Treatment of Cancer of the Tongue.
- D. Radiation Treatment of Cancer of the Cervix.

The last two subjects will cover essentially the following: (1) the technic of radiotherapy, (2) immediate and subsequent reactions, (3) case reports illustrating degree and histologic type of cancer.

The official speakers will be named by the Executive Committee after duly considering the recommendations from the committee of each country. The official speaker may present one or more lectures.

Prospective members from the United States who wish to participate in any of the discussions outlined above should communicate with Dr. Case at their earliest convenience, so that the American committee will be able to adjust the discussions to the program by conference with the Program Committee in Santiago.

In addition to these official presentations, there

will be time set apart for free subjects not related to the official themes. These may be presented only in the scientific exhibits, in order to give them more objectivity. The free subjects will be presented as exhibits with comments.

The Inter-American College of Radiology was organized at the close of the Second Inter-American Congress of Radiology in Havana, Cuba, November, 1946. A convocation of this College will be held in connection with the Congress. Information concerning the College and application blanks for fellowship in it may be obtained from Dr. Case. Membership in the Third Inter-American Congress of Radiology is, however, independent and not contingent upon fellowship in the College.

The more than three hundred Americans who made up the attendance from the United States at the Second Inter-American Congress of Radiology in Havana have very pleasant memories of the meeting, and many of them will wish to attend the Congress in Santiago in 1949. The month of November is probably the most favorable time of the year from the standpoint of weather. It should be borne in mind that the seasons are reversed, our winter being the summer of our South American friends. Excursions are contemplated to the southern part of Chile, especially the lake region, which is very beautiful and which offers opportunities for fishing and skiing, even in the summer months (corresponding to winter in the United States).

There is talk of arranging parties for air and sea travel, including a tour of some of the chief cities of South America. By air travel, a very reasonable tour of the principal cities of South America and attendance on the full period of the Congress can be accomplished in about twenty days. Our Latin-American colleagues will give generously of their time in facilitating visits to the principal radiological institutions of each large city visited in the course of such a tour.

Further notices will be published from time to time. If any are anticipating, even faintly, attendance at this meeting, it is requested that they communicate promptly with Dr. Case. Every endeavor will be made to supply full information.

Address all communications to the chairman of the committee, Dr. James T. Case, 55 East Washington Street, Chicago 2, Ill.

FIRST ANNUAL NATIONAL CANCER CONFERENCE

Approximately 400 U. S. cancer control leaders will meet in Memphis, Tenn., for the first annual National Cancer Conference, Feb. 25-27, 1949.

The Conference will be sponsored jointly by the American Cancer Society and the National Cancer Institute of the Public Health Service, Federal Security Agency. Sessions will be held at the Hotel Peabody.

Various phases of the cancer problem will be dis-

cussed at round-table forums. Participants will include outstanding doctors and scientists from all parts of the country in the fields of medicine, nursing, statistics, research, and public health.

A CORRECTION

In the paper on "Roentgen Treatment of Multiple Myelomatosis" by Garland and Kennedy in the March 1948 issue of RADIOLOGY, reference is made (page 301) to Pohle as favoring irradiation in the treatment of that condition and he is directly quoted as follows: "As myeloma is very radiosensitive, a total dose of 900 to 1,200 r . . . should be a sufficient amount of irradiation. Smaller areas of bone involvement may be treated by a single dose of 600 r."

Actually, though this reference is to Pohle's book (*Clinical Roentgen Therapy*, Lea & Febiger, 1938), it is to a chapter by Otto Jüngling, to whom the views expressed should be credited. Lest the statement be taken as indicating an over-optimistic attitude, it may be mentioned that Jüngling states quite definitely that the prognosis in this disease is poor.

In offering this correction, with their apologies, the authors take the opportunity of adding a note on one of the cases (Case 13) reported in their own paper, with an extraordinarily long survival. This patient died of bronchopneumonia in the summer of 1948. At autopsy he was found to have advanced myelomatosis.

Book Reviews

STANDARDS FOR THE DIAGNOSIS AND TREATMENT OF CANCER. By The Cancer Committee of the Iowa State Medical Society. Second Edition. A volume of 160 pages. Published by the Athens Press, Iowa City, Ia. Price \$1.00.

This cancer manual is addressed primarily to the private medical practitioner, presenting the salient facts regarding early cancer and how to recognize and properly care for it. Simply written, containing a wealth of information regarding cancer as it involves the various parts of the body, this little book could well find a place in the library of every practicing physician.

In Memoriam

NELIUS JULIAN NESSA, M.D.

1880-1948

Dr. Nelius J. Nessa of Sioux Falls, South Dakota, a pioneer in radiology in the Middle West, and the first radiologist to establish a practice in South Dakota, died in Rochester, Minn., on July 2, 1948, of acute leukemia.



Nelius Julian Nessa, M.D.

Dr. Nessa was a native of Minnesota. He was graduated from the University of Minnesota in 1904, was licensed to practice in South Dakota in 1909, and in 1919 associated himself with others in the founding of the Sioux Falls Clinic, with which he continued to be connected until his death. He was also on the staffs of the McKennan and Sioux Valley Hospitals.

Dr. Nessa became a member of the Radiological Society of North America in 1920 and served as Counselor for South Dakota from that year until his

death. He was a diplomate of the American Board of Radiology and a member of the American Roentgen Ray Society, the American College of Radiology, and the Minnesota Radiological Society.

SIDNEY CONROY BARROW, M.D.

1876-1948

Dr. Sidney C. Barrow of Shreveport, La., a member of the Radiological Society of North America for thirty years, died on Aug. 11, 1948. He had suffered an attack of coronary thrombosis several years ago but, in spite of this, continued to carry on his practice until two weeks before his death.

Dr. Barrow was born in Feliciana Parish, La., in 1876. He was graduated from Centenary College in 1896 and obtained his degree in medicine from the University of Tennessee in 1901. In 1906 he moved to Shreveport and there established himself in the practice of radiology, being thus the pioneer radiologist of the state. For a long period he was the only radiologist in Shreveport and did an excellent job of supplying services to the various hospitals and clinics as well as maintaining his private practice. In more recent years he was associated with Dr. W. R. Harwell.

Dr. Barrow was a firm believer in organized medicine, and was held in high regard by his fellow practitioners, serving as president of the State Medical Society and the Shreveport Medical Society. Beside his membership in the Radiological Society of North America, he was a member of the American Roentgen Ray Society and of the American Radium Society and a fellow of the American College of Radiology. He was a frequent contributor to the radiological literature.

Genial, bright, and industrious, Dr. Barrow kept well abreast of the times. His confreres feel keenly the loss of a dear friend and colleague. He is survived by a daughter, Mrs. J. U. Galloway, of Shreveport, and a sister, Mrs. Lois West, of Baton Rouge.

LUCIEN A. FORTIER, M.D.

RADIOLOGICAL SOCIETIES: SECRETARIES AND MEETING DATES

Editor's Note: Secretaries of state and local radiological societies are requested to co-operate in keeping this section up-to-date by notifying the editor promptly of changes in officers and meeting dates.

UNITED STATES

RADIOLOGICAL SOCIETY OF NORTH AMERICA. *Secretary-Treasurer,* Donald S. Childs, M.D., 713 E. Genesee St., Syracuse 2, N. Y.

AMERICAN RADIUM SOCIETY. *Secretary,* Hugh F. Hare, M.D., 605 Commonwealth Ave., Boston 15, Mass.

AMERICAN ROENTGEN RAY SOCIETY. *Secretary,* Harold Dabney Kerr, M.D., Iowa City, Iowa.

AMERICAN COLLEGE OF RADIOLOGY. *Secretary,* William C. Stronach, 20 N. Wacker Dr., Chicago 6, Ill.

SECTION ON RADIOLOGY, A. M. A. *Secretary,* U. V. Portmann, M.D., Cleveland Clinic, Cleveland 6, Ohio.

Alabama

ALABAMA RADIOLOGICAL SOCIETY. *Secretary-Treasurer,* W. D. Anderson, M.D., 2501 6th St., Tuscaloosa.

Arkansas

ARKANSAS RADIOLOGICAL SOCIETY. *Secretary,* Fred Hames, M.D., Pine Bluff. Meets every three months and at meeting of State Medical Society.

California

CALIFORNIA MEDICAL ASSOCIATION, SECTION ON RADIOLOGY. *Secretary,* Sydney F. Thomas, M.D., Palo Alto Clinic, Palo Alto.

LOS ANGELES RADIOLOGICAL SOCIETY. *Secretary,* Moris Horwitz, M.D., 2009 Wilshire Blvd., Los Angeles 5. Meets second Wednesday of each month at County Society Bldg.

NORTHERN CALIFORNIA RADIOLOGICAL CLUB. *Secretary,* Charles E. Grayson, M.D., Medico-Dental Bldg., Sacramento 14. Meets at dinner last Monday of September, November, January, March, and May.

PACIFIC ROENTGEN SOCIETY. *Secretary,* L. Henry Garland, M.D., 450 Sutter St., San Francisco 8. Meets annually with State Medical Association.

SAN DIEGO ROENTGEN SOCIETY. *Secretary,* R. F. Niehaus, M.D., 1831 Fourth Ave., San Diego. Meets first Wednesday of each month.

X-RAY STUDY CLUB OF SAN FRANCISCO. *Secretary,* Ivan J. Miller, M.D., 49 Fourth St. Meets monthly on the third Thursday at 7:45 P.M., January to June at Lane Hall, Stanford University Hospital, and July to December at Langley Porter Clinic, University of California Hospital.

Colorado

DENVER RADIOLOGICAL CLUB. *Secretary,* Mark S. Donovan, M.D., 306 Majestic Bldg., Denver 2. Meets third Friday of each month, at the Colorado School of Medicine and Hospitals.

Connecticut

CONNECTICUT STATE MEDICAL SOCIETY, SECTION ON RADIOLOGY. *Secretary,* Robert M. Lowman, M.D.,

Grace-New Haven Hospital, Grace Unit, New Haven. Meetings bimonthly, second Thursday. **CONNECTICUT VALLEY RADIOLOGICAL SOCIETY.** *Secretary,* Ellwood W. Godfrey, M.D., 1876 Boulevard, W. Hartford. Meets second Friday of October and April.

District of Columbia

RADIOLOGICAL SECTION, DISTRICT OF COLUMBIA MEDICAL SOCIETY. *Secretary,* Alfred A. J. Den, M.D., 1801 K St., N. W., Washington 6. Meets third Thursday, January, March, May, and October, at 8:00 P.M., in Medical Society Auditorium.

Florida

FLORIDA RADIOLOGICAL SOCIETY. *Secretary-Treasurer,* F. K. Hurt, M.D., Riverside Hospital, Jacksonville. Meets in April and in November.

Georgia

GEORGIA RADIOLOGICAL SOCIETY. *Secretary-Treasurer,* Robert Drane, M.D., De Renne Apartments, Savannah. Meets in November and at the annual meeting of State Medical Association.

Illinois

CHICAGO ROENTGEN SOCIETY. *Secretary,* T. J. Wachowski, M.D., 310 Ellis Ave., Wheaton. Meets at the Palmer House, second Thursday of October, November, January, February, March, and April, at 8:00 P.M.

ILLINOIS RADIOLOGICAL SOCIETY. *Secretary-Treasurer,* William DeHollander, M.D., St. Johns' Hospital, Springfield. Meetings quarterly as announced.

ILLINOIS STATE MEDICAL SOCIETY, SECTION ON RADIOLOGY. *Secretary,* John H. Gilmore, M.D., 720 N. Michigan Ave., Chicago 11.

Indiana

INDIANA ROENTGEN SOCIETY. *Secretary-Treasurer,* William M. Lochr, M.D., 712 Hume-Mansur Bldg., Indianapolis 4. Annual meeting in May.

Iowa

IOWA X-RAY CLUB. *Secretary,* Arthur W. Erskine, M.D., 326 Higley Building, Cedar Rapids. Meets during annual session of State Medical Society.

Kansas

KANSAS RADIOLOGICAL SOCIETY. *Secretary-Treasurer,* Anthony F. Rossitto, M.D., Wichita Hospital, Wichita. Meets annually with State Medical Society.

Kentucky

KENTUCKY RADIOLOGICAL SOCIETY. *Secretary-Treasurer,* Sydney E. Johnson, M.D., 101 W. Chestnut St., Louisville.

LOUISVILLE RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, Everett L. Pirkey, Louisville General Hospital, Louisville 2. Meets second Friday of each month at Louisville General Hospital.

Louisiana

LOUISIANA RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, Johnson R. Anderson, M.D., No. Louisiana Sanitarium, Shreveport. Meets with State Medical Society.

ORLEANS PARISH RADIOLOGICAL SOCIETY. *Secretary*, Joseph V. Schlosser, M.D., Charity Hospital of Louisiana, New Orleans 13. Meets first Tuesday of each month.

SHREVEPORT RADIOLOGICAL CLUB. *Secretary*, Oscar O. Jones, M.D., 2622 Greenwood Road. Meets monthly September to May, third Wednesday.

Maryland

BALTIMORE CITY MEDICAL SOCIETY, RADIOLOGICAL SECTION. *Secretary*, J. Howard Franz, M.D., 1127 St. Paul St., Baltimore 2.

Michigan

DETROIT X-RAY AND RADIUM SOCIETY. *Secretary-Treasurer*, E. R. Witwer, M.D., Harper Hospital, Detroit 1. Meetings first Thursday, October to May, at Wayne County Medical Society club rooms.

MICHIGAN ASSOCIATION OF ROENTGENOLOGISTS. *Secretary-Treasurer*, R. B. MacDuff, M.D., 220 Genesee Bank Building, Flint 3.

Minnesota

MINNESOTA RADIOLOGICAL SOCIETY. *Secretary*, C. N. Borman, M.D., 802 Medical Arts Bldg., Minneapolis 2. Meets in Spring and Fall.

Missouri

RADIOLOGICAL SOCIETY OF GREATER KANSAS CITY. *Secretary*, Wm. M. Kitchen, M.D., 1010 Rialto Building, Kansas City, 6, Mo. Meetings last Friday of each month.

ST. LOUIS SOCIETY OF RADIOLOGISTS. *Secretary*, Edwin C. Ernst, M.D., 100 Beaumont Medical Bldg. Meets on fourth Wednesday, October to May.

Nebraska

NEBRASKA RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, Ralph C. Moore, M.D., Nebraska Methodist Hospital, Omaha 3. Meets third Wednesday of each month at 6 P.M. in Omaha or Lincoln.

New England

NEW ENGLAND ROENTGEN RAY SOCIETY. *Secretary-Treasurer*, George Levene, M.D., Massachusetts Memorial Hospitals, Boston. Meets monthly on third Friday at Boston Medical Library.

New Hampshire

NEW HAMPSHIRE ROENTGEN SOCIETY. *Secretary-Treasurer*, Albert C. Johnston, M.D., Elliot Community Hospital, Keene. Meetings quarterly in Concord.

New Jersey

RADIOLOGICAL SOCIETY OF NEW JERSEY. *Secretary*, Raphael Pomeranz, M.D., 31 Lincoln Park, Newark 2. Meetings at Atlantic City at time of State Medical Society and midwinter in Newark.

New York

ASSOCIATED RADIOLOGISTS OF NEW YORK, INC. *Secretary*, William J. Francis, M.D., East Rockaway.

BROOKLYN ROENTGEN RAY SOCIETY. *Secretary-Treasurer*, Abraham H. Levy, M.D., 1354 Carroll St., Bklyn. 13. Meets fourth Tuesday of every month, October to April.

BUFFALO RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, Mario C. Gian, M.D., 610 Niagara St., Buffalo 1. Meetings second Monday, October to May.

CENTRAL NEW YORK ROENTGEN SOCIETY. *Secretary-Treasurer*, Dwight V. Needham, M.D., 608 E. Genesee St., Syracuse 10. Meetings in January, May, and October.

LONG ISLAND RADIOLOGICAL SOCIETY. *Secretary*, Marcus Wiener, M.D., 1430 48th St., Brooklyn 19. Meetings fourth Thursday evening, October to May, at 8:45 P.M., in Kings County Medical Bldg.

NEW YORK ROENTGEN SOCIETY. *Secretary*, Wm. Snow, M.D., 941 Park Ave., New York 28.

QUEENS ROENTGEN RAY SOCIETY. *Secretary*, Jacob E. Goldstein, M.D., 88-29 163rd St., Jamaica 3. Meets fourth Monday of each month.

ROCHESTER ROENTGEN-RAY SOCIETY. *Secretary*, Murray P. George, M.D., 260 Crittenden Blvd. Rochester 7. Meets at Strong Memorial Hospital, third Monday, September through May.

North Carolina

RADIOLOGICAL SOCIETY OF NORTH CAROLINA. *Secretary-Treasurer*, James E. Hemphill, M.D., Professional Bldg., Charlotte 2. Meets in May and October.

North Dakota

NORTH DAKOTA RADIOLOGICAL SOCIETY. *Secretary*, Charles Heilman, M.D., 1338 Second St., N. Fargo.

Ohio

OHIO STATE RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, Carroll Dundon, M.D., 2065 Adelbert Road, Cleveland 6. Next meeting at annual meeting of the State Medical Association.

CENTRAL OHIO RADIOLOGICAL SOCIETY. *Secretary*, Paul D. Meyer, M.D., Grant Hospital, Columbus. Meets second Thursday, October, December, February, April, and June, 6:30 P.M., Seneca Hotel, Columbus.

CINCINNATI RADIOLOGICAL SOCIETY. *Secretary*, Eugene L. Saenger, M.D., 735 Doctors Bldg., Cincinnati 2. Meets last Monday, September to May.

CLEVELAND RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, George L. Sackett, M.D., 10515 Carnegie Ave., Cleveland 6. Meetings at 6:30 P.M. on fourth Monday, October to April, inclusive.

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Oklahoma

OKLAHOMA STATE RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, W. E. Brown, M.D., 21st and Xanthus, Tulsa 4. Meets in October, January, and May.

Oregon

OREGON RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, Boyd Isenhardt, M.D., 214 Medical-Dental Bldg., Portland 5. Meets monthly, on the second Wednesday, at 8:00 P.M., in the library of the University of Oregon Medical School.

Pacific Northwest

PACIFIC NORTHWEST RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, Sydney J. Hawley, M.D., 1320 Madison St., Seattle 4, Wash. Meets annually in May.

Pennsylvania

PENNSYLVANIA RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, James M. Converse, M.D., 416 Pine St., Williamsport 8. Meets annually.

PHILADELPHIA ROENTGEN RAY SOCIETY. *Secretary*, Arthur Finkelstein, M.D., Graduate Hospital, Philadelphia. Meets first Thursday of each month at 8:00 P.M., from October to May in Thomson Hall, College of Physicians, 21 S. 22d St.

PITTSBURGH ROENTGEN SOCIETY. *Secretary-Treasurer*, R. P. Meader, M.D., 4002 Jenkins Arcade, Pittsburgh 22. Meets second Wednesday of each month at 6:30 P.M., October to June.

Rocky Mountain States

ROCKY MOUNTAIN RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, Maurice D. Frazer, M.D., Lincoln Clinic, Lincoln, Nebr.

South Carolina

SOUTH CAROLINA X-RAY SOCIETY. *Secretary-Treasurer*, Robert B. Taft, M.D., 103 Rutledge Ave., Charleston 16.

Tennessee

MEMPHIS ROENTGEN CLUB. Meetings second Tuesday of each month at University Center.

TENNESSEE RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, J. Marsh Frère, M.D., 707 Walnut St., Chattanooga. Meets annually with State Medical Society in April.

Texas

DALLAS-FORT WORTH ROENTGEN STUDY CLUB. *Secretary*, X. R. Hyde, M.D., Medical Arts Bldg., Fort Worth 2. Meetings on third Monday of each month in Dallas in the odd months and in Fort Worth in the even months.

HOUSTON X-RAY CLUB. *Secretary*, Curtis H. Burge, M.D., 3020 San Jacinto, Houston 4. Meetings fourth Monday of each month.

TEXAS RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, R. P. O'Bannon, M.D., 650 Fifth Ave., Fort Worth 4. Next meeting Jan. 7-8, 1949.

Utah

UTAH STATE RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, M. Lowry Allen, M.D., Judge Bldg., Salt Lake City 1. Meets third Wednesday, January, March, May, September, November.

Virginia

VIRGINIA RADIOLOGICAL SOCIETY. *Secretary*, P. B. Parsons, M.D., Norfolk General Hospital, Norfolk 7.

Washington

WASHINGTON STATE RADIOLOGICAL SOCIETY. *Secretary-Treasurer*, Homer V. Hartzell, M.D., 310 Stimson Bldg., Seattle 1. Meetings fourth Monday October through May, at College Club, Seattle.

Wisconsin

MILWAUKEE ROENTGEN RAY SOCIETY. *Secretary-Treasurer*, Theodore J. Pfeffer, M.D., 839 N. Marshall St., Milwaukee 2. Meets monthly on second Monday at the University Club.

RADIOLOGICAL SECTION OF THE WISCONSIN STATE MEDICAL SOCIETY. *Secretary*, S. R. Beatty, M.D., 185 Hazel St., Oshkosh. Two-day meeting in May and one day with State Medical Society, September.

UNIVERSITY OF WISCONSIN RADIOLOGICAL CONFERENCE. Meets first and third Thursdays 4 P.M., September to May, Service Memorial Institute, Madison 6.

Puerto Rico

ASOCIACIÓN PUERTORRIQUEÑA DE RADIOLOGÍA. *Secretary*, Jesús Rivera Otero, M.D., Box 3524, Santurce, Puerto Rico.

CANADA

CANADIAN ASSOCIATION OF RADIOLOGISTS. *Honorary Secretary-Treasurer*, E. M. Crawford, M.D., 2100 Marlowe Ave., Montreal 28, Quebec. Meetings in January and June.

LA SOCIÉTÉ CANADIENNE-FRANÇAISE D'ELECTROLOGIE ET DE RADIOLOGIE MÉDICALES. *General Secretary*, Origène Dufresne, M.D., Institut du Radium, Montreal. Meets third Saturday each month.

CUBA

SOCIEDAD DE RADIOLOGÍA Y FISIOTERAPIA DE CUBA. Offices in Hospital Mercedes, Havana. Meets monthly.

MEXICO

SOCIEDAD MEXICANA DE RADIOLOGÍA Y FISIOTERAPIA. *General Secretary*, Dr. Dionisio Pérez Cosío, Marsella 11, México, D. F. Meetings first Monday of each month.

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ROENTGEN DIAGNOSIS

THE HEAD AND NECK

Hyperostosis Cranii. Torfinn Denstad. *Acta radiol.* 28: 129-138, April 30, 1947.

The author reports three cases which he believes represent a so-called idiopathic cranial hyperostosis, particularly to show that diffuse cranial hyperostosis is not pathognomonic of meningioma and to emphasize the necessity of using all means to eliminate the possibility of an incorrect diagnosis of that condition. The patients were women, aged 67, 40, and 67 years. The affection involves particularly the sphenoidal wings and floor of the anterior cranial fossa, but also invades the facial bones, with osteoma formation in the sinuses. The x-ray picture resembles that of basal meningioma. In none of the author's cases, however, were bone or vascular changes pathognomonic of meningioma demonstrable, nor was there atrophy or erosion of the dorsum sellae. Neither were the histories or the neurologic findings directly indicative of cerebral tumor. Roentgenograms are reproduced.

Herniation of the Cerebral Ventricles. Charles R. Perryman and Eugene P. Pendergrass. *Am. J. Roentgenol.* 59: 27-51, January 1948.

As the ventricles dilate under pressure caused by blocking of the cerebral ventricular system, there are certain relatively weak areas where herniation may develop. These are the anterior and posterior walls of the third ventricle, the posteromedial floor of the lateral ventricle near the origin of the temporal horn, and the superior portion of the fourth ventricle. Except for the anterior wall of the third ventricle, all the above-mentioned areas are in close anatomical proximity. The resultant hernia presents under the tentorium and usually produces a pressure defect on the superior surface of the cerebellum.

The symptomatology arising from the hernia is an unsettled question, since in many cases in which these hernias develop there are already posterior fossa lesions which are themselves producing cerebellar symptoms.

The anatomy and pathological physiology are fully related, and a number of illustrative cases are presented. Most of the roentgenograms are well reproduced, as well as several interesting photographs of the gross specimens.

The treatment is that of the underlying disease, with removal of the sac in cases where only palliation can be achieved, since the presence of a large sac herniating from the third or lateral ventricles beneath the resistant tentorium would surely contribute to blockage of the aqueduct of Sylvius and thus establish a vicious circle.

ZAC F. ENDRESS, M.D.

Diagnostic Significance of Change in Position of Metallic Foreign Bodies in Brain Abscess. Ernest H. Wood, Jr. *Am. J. Roentgenol.* 59: 52-58, January 1948.

Metallic foreign bodies retained in the brain substance following penetrating wounds become valuable indicators of the presence of abscess or hematoma. Since the abscess develops in the necrotic tissue in the path of the missile, the foreign body will be surrounded by the liquefied brain tissue. In films taken in different positions the foreign body will then be seen to change position under the influence of gravity. The same holds

true of a hematoma, since most of the hemorrhage seems to take place at the end of the missile path.

Two cases showing gravitational shift are presented as well as 2 in which there was a change in the position of a fragment in three weeks time although no gravitational shift was demonstrated. It was seen at operation that the fragments were imbedded in the walls of abscess cavities.

This is a valuable sign, well worth remembering, since many of these cases do not give a clear-cut clinical picture of an expanding inflammatory lesion because of the tissue loss along the missile track.

ZAC F. ENDRESS, M.D.

A Huge Calcified Intracranial Oligodendroglioma. C. Eugene DeAngelis and Elizabeth Sammis DeAngelis. *South. M. J.* 40: 838-840, October 1947.

Intracranial calcifications occur in certain normal structures of the brain, as well as in pathological entities. Fifty-nine per cent of adult pineal glands can be visualized in the mid-line on the anteroposterior view of the skull; the falx cerebri may show calcific deposits, particularly in the frontal region; the tentorium cerebelli may be calcified, and lastly the choroid plexuses can often be seen as bilateral intracranial shadows.

Calcific deposits are demonstrable in 41 per cent of suprasellar cysts and in 31 per cent of oligodendrogliomas, 7 per cent of dermoids, and an occasional astrocytoma or chordoma. Reports of large calcified intracranial tumors, however, are rare. Light reported an unusual case in which an almost uniformly dense shadow, 7×7 cm., in the left frontal region proved to be an oligodendroglioma (*Ann. Surg.* 117:309, 1943).

The authors' patient was a fifty-year-old white male who had had mental symptoms for over a year, suggestive of a frontal lobe type of syndrome. Roentgenograms of the skull, which are reproduced in the article, show a large, oval, confluent calcific mass in the right lower and lateral aspect of the parietal lobe, measuring $7.5 \times 4 \times 4.5$ cm. The margins are somewhat serrated, suggesting conformity with the adjacent convolutional configurations. The sella turcica was normal. There was slight calcification in the falx cerebri, which was seen to lie in the mid-line. The calvarium was considered to be normal. A large, stony-hard tumor was successfully removed, measuring approximately 7.5×5 cm. The histologic picture was typical of oligodendroglioma.

WILLIS MANGES, M.D.

Absence of the Anterior Cerebral Artery in the Skull Arteriogram. M. Zehnder. *Schweiz. med. Wchnschr.* 77: 1356-1359, Dec. 27, 1947.

The anterior cerebral artery in the anteroposterior projection shows a lateral displacement under the falx, similar to the ventricular displacement in ventriculography, in the presence of a space-occupying lesion, but a similar defect may also occur in the presence of extracerebral disease (i.e., subdural hematoma). In the lateral projection, extracerebral medial tumors may lead to elevation and inward bulging of the artery. Frontal lobe tumors may also produce pathological patterns in the carotid siphon and the anterior cerebral artery, and Fischer has used this sign to work out an exact system of localization for the surgical attack (*Zentralbl. f. Neurochir.* 4: 72, 1939).

In three cases here reported, all of different diseases, arteriographic study of the involved side showed absence of the anterior cerebral artery. All the patients complained of supraorbital pain, and two had mental changes, originally ascribed to the basic disease. It is not always clear whether the arteriographic anomaly was due to spasm or to anatomical variation. In two of the reported cases (ruptured aneurysm and tumor metastasis to the anterior fossa) anatomical obliteration is probable; in the other case (rhinologic infection) this is less clear. In the first case a control study of the opposite side showed an anomalous doubling of the artery. These cases seem to indicate that the absence of the anterior cerebral artery is a significant finding.

LEWIS G. JACOBS, M.D.

Mucocoele of the Fronto-Ethmoid Area: Report of Two Cases. William D. Morman. *Ann. Otol., Rhin. & Laryng.* 56: 927-931, December 1947.

Two cases of mucocoele of the fronto-ethmoid region are reported. Both of these patients complained of symptoms related to the eyes: loss of vision in one instance and epiphora in the other. In each case physical examination revealed a fluctuant mass. X-ray examination in the first showed the maxillary sinus to be obscured. Following lipiodol injection the medium was seen in the lower portion of a cyst, with a fluid level above it. In the second case an opacity involving the right frontal, ethmoid, and both maxillary sinuses was demonstrable. In neither of these cases was the x-ray study of diagnostic value until operation was undertaken and cystic masses containing thick brown or yellowish secretion were discovered. The author believes, nevertheless, that all patients with pressure symptoms around the inner canthus should be examined roentgenologically for possible ethmoid sinus involvement. The pathologist's report, after extensive removal of the sac walls in the cases reported, was chronic inflammation.

In the earlier stages of mucocoele involving the frontal and ethmoid sinuses, all symptoms may be lacking, as the growth usually takes place painlessly. Not until marked swelling occurs does the patient pay any attention to this condition. During the later stages, a hard, bony distention may be encountered that may confuse the clinician by its similarity to malignant growth. A parchment-like crackling due to partial absorption of the bone should lead one to suspect a mucocoele.

STEPHEN N. TAGER, M.D.

Wooden Foreign Body in the Ethmoid Capsule. Alfred J. Cantoni. *Ann. Otol., Rhin. & Laryng.* 56: 953-956, December 1947.

Roentgen identification of a non-metallic foreign body is often difficult. The author reports the case of a Negro male who had been stabbed in the left conjunctival sac with a common wooden pencil four months before examination. Anterior rhinoscopy disclosed an elongated mass firmly embedded between the left lateral nasal wall and the septum, at the level of the inferior border of the middle turbinate, bathed in mucopus. Under general anesthesia, two portions of a wooden pencil were removed through the anterior nares, one piece, measuring 3.5 cm., from the middle third of the left meatus, the other, measuring 3.75 cm., from the posterior third of the right nasal cavity. All of the graphite was accounted for except about 3.5 cm. from the pointed end of the pencil.

Films obtained prior to operation had shown only clouding of the sinuses and some mucosal thickening. On re-examination of these films, however, there was apparent in the lateral view a slight increase in density approximately 6 cm. in length and less than 3 mm. in diameter extending from the floor of the orbit obliquely downward and posteriorly to the floor of the nasal cavity. A lateral film taken subsequent to operation showed a similar shadow about 3 cm. in length. This apparently represented the missing portion of graphite, which later was found extruded on the floor of the right nasal cavity adherent to dry mucopurulent material. X-ray examination thereafter was normal.

STEPHEN N. TAGER, M.D.

Calcified Hematoma of the Oropharynx Secondary to a Gunshot Wound. Edward H. Diamond and Charles W. Perkins. *Arch. Otolaryng.* 47: 64-66, January 1948.

A 70-year-old retired policeman first sought medical advice in August 1946 because of progressive impairment of hearing in the left ear. There was tinnitus in this ear occasionally, and intermittent supraorbital pain on the left side. In 1917 the patient had been shot during the performance of duty, the bullet traversing the left malar region and lodging near the body of the second cervical vertebra. Following the injury he was semiconscious for a time and had what was apparently a partial left hemiplegia. The bullet was left *in situ* and there had been no symptoms since that time except some soreness of the left parietal region and occasional pain in the left ear.

The tympanic membrane appeared normal on examination. Hearing (whispered voice) was 15/15 in both ears. The left eustachian tube did not inflate readily. The left lateral posterior pharyngeal wall presented a large hemispheric swelling about 4 cm. in diameter, smooth and bony hard.

Films taken in 1934 showed that the bullet passed from the left side of the nose through the roof of the mouth to the atlas-axis region. Films taken in 1946 showed a large calcareous deposit on the left side of the pharynx and vertebral region, obscuring the bullet.

On aspiration of the tumor, 4 c.c. of jelly-like material were obtained; the pathologic report on this was "calcific material." The mass seemed to diminish in size, but roentgenograms taken subsequent to this procedure showed no change. The consensus of opinion was that the mass was a calcified hematoma, not a true neoplasm, and that, unless more serious symptoms developed, no further treatment was indicated.

THE CHEST

Airblock in the Newborn Period. George W. Salmon. *New Orleans M. & S. J.* 100: 253-258, December 1947.

Aberrant air escaping from the respiratory passages as a cause of death in the newborn is often not disclosed at necropsy. It is easily overlooked or, if discovered, may not be considered of sufficient importance.

Experimental and clinical evidence indicates that escape of air from the respiratory passages is into the lung itself, producing pulmonary interstitial emphysema. Mediastinal emphysema and pneumothorax may subsequently occur. Clinical conditions which enhance the development of pulmonary interstitial emphysema are atelectasis, congenital heart disease, intra-

cranial hemorrhage, vigorous attempts at resuscitation and aspiration of mucus, meconium, or amniotic fluid. The symptoms produced by pulmonary interstitial emphysema alone are not certain. When mediastinal emphysema and/or pneumothorax are also present, cyanosis and dyspnea occur.

The roentgen diagnosis of pulmonary emphysema offers little difficulty, although differentiation of compensatory from obstructive emphysema, in infants, may not be easy. Moreover, once the diagnosis of obstructive emphysema is established, it may be impossible to decide whether the major portion is in or outside of the normal passageways. Since interstitial emphysema may complicate both the obstructive and compensatory types, it should be suggested by the sudden onset of any marked emphysema.

The x-ray diagnosis of mediastinal emphysema is not simple. The frontal projections of the chest may reveal curvilinear collections of air along the margins of the mediastinum or heart. The lateral views may show large collections of air in the anterior mediastinum. None of these views is reliable, however, and pneumothorax complicating the picture may negate the value of the lateral view. Severe pulmonary emphysema of the anterior portion of the lung may simulate mediastinal emphysema in the lateral projection.

The roentgen features of pneumothorax are well known. However, there is no relation of the size of the pneumothorax to its pressure. A comparatively small pneumothorax may be under great pressure. If the underlying lung contains sufficient interstitial air, it splints the lung in an expanded position and, being trapped, prevents more than moderate collapse. Similarly, if the opposite lung is involved, it may prevent mediastinal shift, although evidence of a tension pneumothorax is present.

Treatment of airblock in the infant is discussed.

LOUIS BERNSTEIN, M.D.

Nocardiosis. *Nocardia asteroides* Infection Simulating Pulmonary Tuberculosis. Robert P. Glover, Wallace E. Herrell, Fordyce R. Heilman, and Karl H. Pfuetze. *J. A. M. A.* 136: 172-175, Jan. 17, 1948.

Nocardia asteroides is an acid-fast actinomycete which should not be confused with the more widely recognized organism *Actinomyces bovis*. Nocardiosis is a rare disease, but should be kept in mind as a remote etiologic possibility in the differential diagnosis of obscure pulmonary infections.

The authors report a case simulating pulmonary tuberculosis, in which complete recovery appears to have followed intensive sulfadiazine therapy after thorough trials of penicillin and streptomycin treatment proved ineffective.

JOHN F. HOLT, M.D.
(University of Michigan)

Changes in the Lungs in 126 Cases of Asbestosis Observed in Finland. Carl Wegelius. *Acta radiol.* 28: 139-152, April 30, 1947.

Asbestosis of the lungs is an industrial disease not often encountered in the Scandinavian countries, the only 5 cases reported having occurred in Norway. In Finland, however, 126 roentgenologically demonstrated cases of asbestosis were found among 476 workers during the past three years; 34 of this number were employed in the dressing works at the mine and 92 in the factory for finished products.

Saupe's method of classification of asbestosis according to the stage of development (*Arch. f. Gewerbepath. u. Gewerbehyg.* 9: 391, 1939) is used:

The ante-primary stage: Uncharacteristic strengthening of the lung markings, mainly in the middle and basal fields; fine ray shadows and quite slight indications of small nodules in the costophrenic or cardiophrenic angles.

Stage I: Extremely fine network in the middle and basal fields of the lung picture, which is already layered with very small soft spots.

Stage II: Denser lung picture; more numerous nodule shadows, extremely small; beginning decrease in clarity of the middle and basal fields, more especially of the areas close to the heart; hila broad and dense, in general without calcium layering.

Stage III: Marked shadowing of the middle and basal fields in contrast to increased ray-permeable emphysematic apical fields. The contours of the heart are difficult to differentiate against the densified lung tissue. The hila are broader, but not especially dense.

Borderline cases are designated as I-II and II-III.

In most of the 126 cases, roentgen examination has been repeated once a year during the past three years. At the last observation, 94 (75 per cent) of the cases were in the milder stages (I and I-II); 23 (18 per cent) in the medium stages (II and II-III); and 9 (7 per cent) in the most advanced stage (III). The average mean exposure in the three groups was 3.4 years, 7.9 years, and 11.5 years, respectively. During the two-year period of observation, 27 of the 36 cases in the Ante-primary Stage progressed to Stage I, 4 of the 32 cases in Stage I to the intermediate Stage I-II, while the same number passed to Stage II. Two out of 10 Stage II cases advanced to the intermediate Stage II-III; no progression to Stage III was observed. After the beginning of the examinations, 2 known deaths attributable to asbestosis occurred.

In the author's opinion, fluoroscopy adds little information to that obtainable from the roentgenogram in asbestosis. The former procedure was not diagnostic in the 36 cases in the Ante-primary Stage. In 64 cases, Stages I and I-II, the diagnosis "increased marking in the basal fields" could be made fluoroscopically in only 16 patients; in Stages II, II-III, and III perceptible changes could certainly be observed in all cases in which fluoroscopy was employed, but no closer classification was possible. The procedure is of value for the determination of conditions connected with mobility of the thoracic organs, particularly in emphysema and pleural adhesions.

Roentgenograms of cases in the various stages are reproduced.

Transitory Focal Pulmonary Edema and Eosinophilia (Löfller's Syndrome). Arthur T. Henderson and Carleton B. Peirce. *Am. J. Roentgenol.* 58: 391-406, October 1947.

A case is described, in some detail, of transitory shifting lung shadows associated with pronounced fluctuating blood eosinophilia (Löfller's syndrome) in a patient with a personal and family history of allergy. In addition to house dust and ragweed pollen, the patient showed evidence of bacterial allergy to the infecting organism, *Haemophilus influenzae*, a constant finding in the sputum and infected sinuses. Specific vaccine therapy, followed by desensitization with ragweed pollen extract and house dust, was the chief re-

medial measure employed, and the patient made fairly steady improvement, with a gain of 40 pounds in weight, from February 1941 until the Fall of 1942. At that time an exacerbation occurred and *H. influenzae* again appeared in the sputum. The administration of fresh vaccine of this organism was followed almost immediately by joint and muscle pains, fever, and marked purpura of the Schönlein type. During this episode, a transitory bundle branch block lesion was revealed electrocardiographically. The patient was able to resume work in May 1943 and has remained fairly well.

This case presents indisputable evidence that the Loeffler shadows are due to allergic edema of the interalveolar pulmonary tissue with its widespread capillary connections.

Roentgenographic Appearance of Persistent Pulmonary Infiltrates Associated with Sensitivity to Histoplasmin. Michael L. Furcolow, Herbert L. Mantz, and Ira Lewis. Pub. Health Rep. 62: 1711-1718, Dec. 5, 1947.

The authors briefly review the literature on histoplasmosis and go on to point out that proof of the hypothesis that *Histoplasma* is a responsible organism for producing pulmonary infiltrates and calcifications is for the greater part inferential but, on the other hand, is undeniable.

The present paper describes and illustrates the roentgenologic characteristics of persistent pulmonary infiltrations in persons who are sensitive to histoplasmin but not to tuberculin. Among 72 asymptomatic cases in school children in Kansas City, Mo. (see also High, Zwerling, and Furcolow: Pub. Health Rep. 62: 20, 1947. Abst. in Radiology 49: 742, 1947), only a few were limited to the lymph nodes, without associated parenchymal involvement; a few were of the disseminated type, consisting of widely scattered lesions of varying size; approximately two-thirds were nodular, sharply circumscribed foci. The remainder, about a fourth of the total, were diffuse patchy infiltrations with poorly defined borders, which sometimes developed into nodular lesions. All of these types are shown in the beautifully reproduced illustrations which make up the bulk of the paper.

One of the most interesting cases illustrated is that of a fourteen-year-old boy who had disseminated lesions, barely visible roentgenographically, scattered through both lungs. A film made eight years later shows clearly diffuse bilateral miliary calcifications. Excellent illustrations are also furnished of the "halo calcification," in which a central core of calcification is observed. These lesions tend to calcify slowly, and may persist without complete calcification over a considerable period.

The great value of this article lies in the illustrations. It makes one aware that there is one more condition to be considered when isolated pulmonary infiltrates, calcification, or widespread disease appears in the lungs.

SYDNEY F. THOMAS, M.D.

Primary Atypical Pneumonia. Report of Eight Cases with Autopsies. Frederic Parker, Jr., Leslie S. Joffile, and Maxwell Finland. Arch. Path. 44: 581-608, December 1947.

Eight cases of primary atypical ("viral") pneumonia in which autopsies were performed are reported. Clinically these cases were characterized by increasing symptoms of respiratory embarrassment, diffuse moist

râles, and transient areas of atelectasis but no definite signs of pulmonary consolidation. Roentgenographically, there was an extensive miliary soft nodular type of density in the lungs. The serum contained cold agglutinins. Two of the cases were complicated by acute hemolytic anemia, 2 others by severe erythema multiforme exudativum, and 1 by rheumatic heart disease with mitral stenosis.

The characteristic findings on gross examination of the lungs were a congested appearance of the cut surfaces, which were studded with small grayish or dark nodules, and a hyperemia of the mucosa of the trachea and of the bronchi. Histologically, there were a mononuclear type of alveolar exudate, an interstitial infiltration predominantly of plasma cells, and swelling and proliferation of the alveolar lining cells. Bacterial infection played a minor role except in 2 cases, in which there was some abscess formation.

Miniature Chest X-rays. E. T. Fralick, H. T. McClintock, E. P. Snider, and C. A. Wicks. Canad. M. A. J. 57: 567-572, December 1947.

The authors present a study and a brief review of the principles involved in the use of miniature chest films for detecting persons with possible intrathoracic disease. The features that are discussed in some detail are (1) accuracy for screening, (2) usefulness, (3) limitations, and (4) results obtained.

It is felt that photofluorography compares favorably in sensitivity with 14 × 17 radiographs for screening purposes when the fluorographs are studied by skillful radiologists. In the authors' experience 2 per cent of the patients receiving miniature chest films required further examination for possible tuberculous infection. Out of that group approximately 30 per cent proved to have no demonstrable disease. Overcautious interpretation and what the authors call "over-reading" may result in recalling for further study a fair percentage of patients who prove to have entirely normal chests. This is particularly apt to occur when the older age groups are being screened.

Among 126,089 miniature chest films taken in 1946 by the Gage Institute (Toronto), 0.91 per cent disclosed tuberculous disease—0.11 per cent active and .80 per cent inactive—confirmed by subsequent examination in the clinic; 0.61 per cent showed evidence of significant non-tuberculous disease or other abnormalities; 1.9 per cent showed less significant conditions, such as previous pleurisy or pulmonary calcifications; in 96.4 per cent no disease of any sort was demonstrable.

In conclusion, the authors mention briefly some of the problems that remain to be solved in utilization of the screening technic as it affects different groups, as hospital admissions, outpatients, food handlers, pre-employment personnel, etc. WILLIS MANGES, M.D.

Mass X-ray Survey in Burlington County [New Jersey]. Alexander Krasnitz. J. M. Soc. New Jersey 44: 493-495, December 1947.

During 1946 a mass chest x-ray survey was conducted in Burlington County, New Jersey, with 70-mm. film and a mobile photofluorographic unit. A total of 4,553 films were read. In 55 (1.2 per cent) there was either definite evidence of tuberculosis or the findings were suspicious. The actual incidence of tuberculosis is unknown, since 20 of the group refused re-examination with 14 × 17-inch films. Sixteen

of the 55 cases were definitely tuberculosis; 4 patients of this group had old fibrotic lesions which were thought to be inactive. Of the 39 "suspicious" cases, 8 were known to the local Tuberculosis League.

One hundred and fifty-six cases of cardiac conditions worthy of notice were recorded, also 51 cases of pleurisy, about one-third of which were assumed to be active.

The author believes the 70-mm. films to be good for screening purposes only and not for diagnosis or for detailed study. In the few instances in which 70-mm. and 14 X 17-inch films were both available, there was a disagreement in interpretation of 17 per cent.

Georgia's Community-Wide Surveys: Chest X-Ray, Blood Test and Dental. Lester M. Petrie. South. M. J. 40: 821-825, October 1947.

Since October 1945 community-wide health surveys have been made in the Savannah, Columbus, Macon, and Augusta areas in Georgia. A total of 215,000 persons out of an estimated population of 438,000 persons of all ages were examined by x-ray and blood tests. The majority were over twelve years of age. The entire program was voluntary and the x-ray survey was the first to be done on a large scale in this area. The author is very enthusiastic about the response of the population, both Negro and white. He feels that the combination of blood and x-ray studies added to the effectiveness of both venereal disease and tuberculosis case finding.

The chest films were made with the 70-mm. photo-fluorographic unit. Besides their value in the detection of tuberculosis, the films brought to light many unrecognized conditions of the heart, mediastinum, and lung fields that otherwise would have gone uncareed for. The results of all the findings, as far as individual patients were concerned, were purely a confidential matter, and the patient was given advice as where to seek the best medical aid for his particular problem. The blood serology test was done by the Mazzini rapid slide precipitation method. Confirmatory studies, including Kahn tests, were made in all cases that were suspected of being positive.

A statistical analysis of several groups of patients under study is given, and some aspects of the public-health problems are discussed. The author believes that the survey methods are of considerable value and that the combined mass survey technic was more effective than any previous case-finding procedure.

WILLIS MANGES, M.D.

Congenital Cystic Disease of the Lung (A Familial Incidence of Four Males in One Family). D. F. McRae. Canad. M. J. 57: 545-550, December 1947.

Four cases of cystic disease of the lung occurring in one family are presented. The author reviewed the literature and found the cases unique in their familial incidence, in the presence of extensive clubbing of the fingers and toes without pulmonary sepsis, and in the malignant nature of the disease, which resulted in the death of all four patients before adult life was reached.

Roentgenograms from two of the cases are reproduced, illustrating the gradual and steady progress of the disease. Involvement appeared most severe at the apices, with gradual diminution toward the bases, an observation confirmed at autopsy. In only one of the cases was an adequate microscopic study made of the lungs. This showed numerous cystic spaces with

fibrosis and some round-cell infiltration of the intervening tissues.

Most investigators feel that a congenital defect is the basis for cystic disease of the lung. Some workers who accept this theory believe that measles is a potent contributory factor, but this was not found to be true in the four cases presented. The cyanosis and pallor in congenital cystic disease are attributed to the cutting down of the pulmonary circulation by the fibrous tissue changes in the lungs rather than to anemia.

JOHN DECARLO, M.D.

Fibroma of the Trachea. John S. Knight and William P. Bunting. Arch. Otolaryng. 47: 67-70, January 1948.

The first case of fibroma of the trachea was discovered at autopsy in 1767 and since that time only 35 tumors of this type have been recorded. An additional case is reported by the author. The patient was a 20-year-old male student, whose chief complaints were bloody sputum, dyspnea, and wheezing. A lateral cervical roentgenogram revealed a large mass protruding from the posterior wall of the trachea, 2 inches below the larynx. There appeared to be almost complete occlusion of the tracheal lumen. Adenoma of the trachea was considered to be the most likely diagnosis. Bronchoscopy showed a large, lobulated reddish mass with intact mucosa about 2 inches below the level of the vocal cords. The basal attachment to the posterior tracheal wall was firm and at least a half inch in diameter. The mucosa was continuous between the tumor and the tracheal wall. Because of the extensive size, the broad base, and the danger of hemorrhage, the tumor was removed by external incision. The histologic diagnosis was soft fibroma of the trachea. Convalescence was uneventful.

The Heart in Normal Infants and Children. Incidence of Precordial Systolic Murmurs and Fluoroscopic and Electrocardiographic Studies. Nathan Epstein. J. Pediat. 32: 39-45, January 1948.

The author studied 280 children for an average of five years, examining each at least four times annually, with electrocardiography and fluoroscopy at least once a year. His observations are in accord with the recognized frequency of benign precordial systolic murmurs of unknown origin. At least 50 per cent of his patients had systolic murmurs in the absence of organic disease. Over half of these patients had murmurs indistinguishable from that of mitral insufficiency. The cardiac shadow was examined in the postero-anterior and both oblique views. No abnormalities were noted. The electrocardiograms revealed the P-R interval to be 0.14 ± 0.02 in the first three years of life, with slightly lower values in later years. Of importance were the unusual and aberrant electrocardiographic findings in 18 per cent of these "normal" children, emphasizing the need for caution in electrocardiographic interpretation.

PAUL W. ROMAN, M.D.

Roentgen Demonstration of Calcifications in the Interventricular Septum in Cases of Heart Block. Frank Windholz and Charles Grayson. Am. J. Roentgenol. 58: 411-421, October 1947.

Anatomical and pathological fundamentals of calcification in the membranaceous septum of the heart are discussed and the technic of their roentgen localization and identification is described. The findings in 49

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patients with annular and valvular calcifications with-
out heart block were compared with those in 12 similar
cases with heart block.

Three types of roentgen signs are considered as indica-
tive of the presence of calcareous deposits in the mem-
branaceous septum: (a) caudad extension of calcium
shadows from calcified aortic valves or aortic ring; (b)
complete circular or crescent-shaped calcifications about
the mitral ostium; (c) incomplete mitral ring calcifica-
tions with nodular thickenings of calcareous deposits at
the right (medial) end of the posterior branch of the
calcified mitral ring.

Calcium deposits in the septum seen on the roentgeno-
gram are as a rule associated with heart block or pro-
longed conduction time, though at times septal calci-
fications can be demonstrated roentgenologically when
clinical and electrocardiographic signs of heart block are
absent.

Clinically Primary Tuberculous Pericarditis. Theo-
dore R. Stepmann and Edwin Owyang. *Ann. Int. Med.*
27: 914-922, December 1947.

The "clinically primary" type of tuberculous peri-
carditis is defined as that in which, at the onset of symp-
toms, there are no clinically demonstrable active
tuberculous lesions elsewhere in the body although at
some later date active foci may be found. This does
not preclude the presence of hidden active foci at the
onset. In distinction, "primary tuberculosis" of the
pericardium, as defined by Thompson, is that in which,
as far as can be ascertained at autopsy, the pericardial
involvement was either the only or the oldest tubercu-
lous lesion.

A total of 50 cases believed to be "clinically primary"
tuberculous pericarditis has been collected from the
literature, to which the authors add 3 more. This paper
concerns itself with a discussion of several unusual
features in these 3 cases as well as an analysis of the
symptoms and signs of those reviewed.

The authors' first patient tolerated a relatively large
pericardial effusion for approximately twenty-four
months. This they explain as due to the fact that the
accumulation of fluid in the pericardial cavity is suf-
ficiently slow to allow the pericardium to accommodate
itself to the increased distention, thereby allowing the
intrapericardial pressure to remain below the level which
causes serious embarrassment to the circulation.
Pneumopericardium was instituted in this case. This
procedure is helpful in both diagnosis and treatment.
Upon production of a pneumopericardium, the findings
of a normal-sized heart, a thickened pericardium, and
the absence of murmurs point strongly to tuberculosis
as the causative factor. In treatment the procedure
offers the possibility of limiting the tuberculous exudate
to the pericardial cavity, thereby averting a later ad-
hesive pericarditis. There are those, however, who
doubt its value, believing that it may increase fibrosis.

In the second case, the interesting findings were a
negative tuberculin skin test (1.0 mg. of old tuberculin)
and the rapid accumulation of fluid, requiring frequent
removal from both pericardial and pleural cavities for
relief.

In the third case, although acid-fast bacilli were not
found in the pericardial fluid, the clinical picture was
adequate for the diagnosis of tuberculous pericarditis.
Tubercle bacilli were subsequently found in the
gastric contents.

Nearly all of the reported cases (31 of 37 for which

adequate data were available) had a fatal outcome.
The duration in the majority of cases was less than six
months. The authors' first case, with a lapse of five
years from onset to death from miliary tuberculosis,
represents the longest survival recorded.

Involvement of the pericardium as the first clinical
manifestation of active tuberculosis is not rare. The
diagnosis should be seriously considered under the
following conditions: (a) presenting symptoms of un-
explained fever, shortness of breath, cough, weakness and
chest pain; (b) evidence of pericardial involvement, *vis.*,
friction rub, effusion and characteristic abnormalities in
the roentgenogram; (c) male patient over thirty years
of age, and of the colored race. Failure to demonstrate
the tubercle bacillus does not necessarily rule out the
clinical diagnosis, as shown by the fact that almost half
of the cases in the series analyzed required postmortem
confirmation.

STEPHEN N. TAGER, M.D.

Calcification of the Myocardium. L. Minor Black-
ford. *Ann. Int. Med.* 27: 1036-1040, December 1947.

A man 59 years of age experienced massive myocardial
infarction on May 4, 1936. He was able to resume his
work the following September and continued with
apparently no cardiac symptoms until about nine years
later. He died in January 1946. At autopsy, the
anterior surface of the left ventricle and apex felt like
bone. The pericardium was plastered against this
portion of the heart, and there were adhesions, also, be-
tween the heart and the diaphragmatic surface of the
pericardium. The anterior descending branch of the
left coronary was obliterated about 2 cm. below the bi-
furcation. Distal to this the heart was ballooned out
and largely calcified. The infarcted area was roughly
circular, 8 cm. in diameter, involving the apex, the an-
terior part of the left ventricle, and about 1 cm. of the
intraventricular septum.

In 1939 a fluoroscopic examination had shown moder-
ate enlargement of the heart, with aneurysm of the left
ventricle, and a tortuous, slightly dilated aorta. An
electrocardiogram was then interpreted as indicating an
old infarction of the anterior wall with left bundle
branch block. Fluoroscopy was again done in Novem-
ber 1945, about six months after the recurrence of symp-
toms, but hurriedly because of the severe dyspnea. It
is believed that a more prolonged fluoroscopic examina-
tion at that time, or an antemortem film, would have
revealed the massive calcification of the myocardium.

The differential diagnosis involves chiefly calcifica-
tion of the pericardium with possible extension into the
myocardium. It is important to make the distinction,
for if the calcification is limited to the pericardium and
symptoms and signs of constriction are present, opera-
tive procedures may effect a cure.

STEPHEN N. TAGER, M.D.

Cardiac Findings Due to Sternal Depression. Report
of Two Cases. Allan E. Moe. *Minnesota Med.* 30:
1265-1267, December 1947.

The author presents two cases in which he attributes
abnormal cardiac findings to sternal depression. The
abstractor cannot agree to this. Illustrations of the
first case show a typical rheumatic heart, in spite of a
mass of laboratory data which seemed to the author to
rule out any suspicion of cardiac disease. The second
case is more doubtful, but the roentgenograms show the
left ventricle at least on the edge of being disproportion-

tionately large. It would be interesting to follow this patient for a few years. PERCY J. DELANO, M.D.

A New Anomaly of the Aorta. Left Aortic Arch with Right Descending Aorta. Raphael N. Paul. *J. Pediatr.* 32: 19-29, January 1948.

A classification of anomalies of the aorta is presented and the common ones are reviewed. A new anomaly, left aortic arch with a right descending aorta, is described for the first time in the literature, and two cases with characteristic x-ray findings are reported. Both patients were originally considered to have a tetralogy of Fallot. The anomaly in each case was diagnosed preoperatively and proved at operation. The diagnosis was based on the following roentgenographic findings:

1. Esophagus, visualized fluoroscopically following routine administration of barium, was indented on the left by the arch of the aorta.
2. There was an absence of any vascular or cardiac shadow to the left of the barium column for a short distance between the aortic knob and the upper left border of the heart.
3. The esophagus descended far to the left of the mid-line, suggesting that the esophagus coursed to the left of the descending aorta.
4. The left anterior oblique view showed anterior displacement of the barium-filled esophagus due to the retro-esophageal aorta emerging diagonally on the right.

Excellent diagrams and photographs are presented to illustrate the diagnostic features listed above.

STANLEY H. MACHT, M.D.

THE DIGESTIVE SYSTEM

Certain Roentgen Manifestations of Gastric Lesions. R. A. Carter and J. E. Vickers. *California Med.* 67: 363-367, December 1947.

The niche of typical benign ulcer has one of the most "characteristic" appearances encountered in gastrointestinal roentgenology. The smooth-margined protrusion from a usually broader base, ending in a rounded, flat, or bluntly conical floor, and the apparent projection of the niche beyond the confines of the stomach are classical. It is almost impossible, however, to make an outright statement that such a lesion is not malignant, and this despite the fact that the size of the ulcer may decrease under a medical regime.

Criteria for malignant ulcers are more reliable: the meniscus sign, irregularity of walls and floor, location (prepyloric and on the greater curvature), and, in general, exceptional size. It is also important that the niche itself remain within the projected outline of the stomach.

Leiomyosarcomas and fibrosarcomas show a central niche. These lesions usually produce a spheroidal tumor not materially deforming the adjacent stomach or impeding its peristalsis. Isolated spheroidal lesions usually appear as negative shadows in a region of the stomach otherwise unmodified as to contour, mucosal pattern, or peristaltic activity. They represent, as a rule, though not invariably, benign tumors or polyps. Irregularity of contour, changes in the adjacent gastric wall, and a niche or button of central necrosis suggest malignancy. Multilobular deformities may be due to polyposis, carcinoma, or sarcoma. Gastritis may present a similar picture.

Gastric retention with dilatation may be due to duodenal ulcer with stenosis and scarring, or to carcinoma of the pylorus.

Prepyloric deformities present a special diagnostic problem. They may be the result of ulcer, gastritis, spasm, benign tumor, polyposis, or extrinsic pressure. Flexibility of the stomach wall, peristalsis, mucosal pattern, presence or absence of a mass are some of the differential points in determining the type of lesion.

Deformities of the cardia may be produced by an intragastric or extragastric tumor. Observation of the gas bubble, mucosal pattern, shape, and flexibility will help in arriving at a correct diagnosis.

The authors emphasize the importance of painstaking and repeated examinations by well trained radiologists. Having gone as far as possible, the radiologist is obligated to present and evaluate the evidence somewhat conservatively, drawing the warranted conclusions but not being led by his desire for a diagnosis to press his evidence to more definite conclusions than it actually supports.

MAURICE D. SACHS, M.D.

Hypertrophic Pyloric Stenosis. Arthur A. Schaefer and John Erbes. *Surg., Gynec. & Obst.* 86: 45-53, January 1948.

Pyloric stenosis is the most common condition requiring surgical treatment in the first few months of life. From June 1924 to May 1947, there were seen in the Milwaukee Children's Hospital 248 cases, 1 for every 213 admissions. There were 9 instances in which more than one member of the family was affected and 7 in which one of twins had the disease. In 72 cases in which operation was performed, microscopic study indicated that the longitudinal as well as the circular smooth muscle fibers were hypertrophied.

The cardinal symptoms of pyloric stenosis are projectile vomiting, failure to gain weight, constipation, visible peristaltic waves, and a palpable pyloric tumor. In the series reported, 77 per cent of the patients had a history of projectile vomiting on entrance to the hospital; 62 per cent were below birth weight; 75 per cent had a history of scanty dark starvation stools; visible peristaltic waves were noted in 83 per cent, and there was a palpable tumor in 63 per cent.

A palpable tumor is considered to be pathognomonic of pyloric stenosis. In 9 cases, however, in which a tumor was palpated, subsequent roentgen study or operation revealed a normal pylorus.

Roentgen examination, properly done, is almost 100 per cent reliable as a diagnostic measure. The pyloric tumor containing barium produces a typical shadow showing the funnel-shaped entrance of the stenosed canal and the long narrow lumen. Delayed opening time and gastric retention of barium are also helpful signs.

The roentgen procedure is as follows: Food is withheld six hours to insure that the stomach is empty. The patient is then given 2 ounces of formula-barium mixture and is observed fluoroscopically. Roentgenograms are taken at five minutes and thirty minutes in the right oblique projection, additional views bring obtained at one and two hours if necessary. The danger of aspiration of barium is considered to be no greater than that of aspiration of vomitus from the periodic full-formula feeding recommended during the preoperative period. In all of the authors' cases lavage was done after roentgen examination and no difficulty was encountered from residual barium in the stomach.

The authors discuss at some length the operative treatment and the preoperative and postoperative management. It is believed that the greatest aid in the reduction of operative mortality in the past ten years has been early diagnosis and adequate preparation.

In the Milwaukee Hospital series, 232 patients were treated surgically, with 9 deaths, an operative mortality rate of 3.8 per cent. Fifteen patients received medical treatment, and of these 7 died, or 46.6 per cent. There has been no death during the past ten years and 172 patients have been operated on without a fatality.

WILLIAM R. ALLEN, M.D.

Emphysematous Gastritis. Claude E. Welch and Chester M. Jones. *New England J. Med.* 237: 983-985, Dec. 25, 1947.

The authors record the fourth case of emphysematous gastritis to appear in the literature. The three previous cases all terminated fatally.

The patient was a 15-year-old girl who experienced severe epigastric cramps at night, following a dinner of fresh salmon. Nausea, vomiting, and diarrhea followed. At first the patient was thought to have an acute gastroenteritis, but the condition persisted and she was admitted to a hospital, where 2,000 c.c. of coffee-ground fluid was evacuated from her stomach and she became quite toxic and shocked. Saline and blood transfusions were administered and she rallied. Six days later x-ray examination showed the stomach to be dilated with a large amount of gas. There were numerous small gas bubbles throughout the fundus and extending down to the antrum, but there was no free air in the peritoneal cavity. A barium meal study showed the gas to be within the gastric wall and it was definitely known that the stomach was empty of food particles. No ulceration was demonstrated within the stomach or duodenum. Culture of the gastric contents showed *Escherichia coli*, non-hemolytic streptococci, and *Staphylococcus aureus*.

It is believed that this patient had an acute gastroenteritis at first, with profuse bleeding and gastric dilatation, at which time the gastric wall was invaded by a combination of the cultured organisms, which produced the gas. Recovery without complications followed penicillin therapy.

Emphysematous gastritis must be differentiated from phlegmonous gastritis and cystic pneumatosis.

JOHN B. McANENY, M.D.

Carcinoma of the Stomach. Symposium. I. A Radiologist's Point of View. J. L. A. Grout. II. **Gastroscopy in the Diagnosis of Gastric Cancer.** H. W. Rodgers. III. **Cancer of the Stomach: Some Pathological Considerations.** Matthew J. Stewart. IV. **The Surgical Aspect of Carcinoma of the Stomach.** Hermon Taylor. V. **Irradiation of Gastric Cancer.** G. Cranston Fairchild and Alan Shorter. VI. **The Statistical Approach to Gastric Cancer.** Denys Jennings. *Brit. J. Radiol.* 20: 491-527, December 1947.

I. Grout points out that the duration of symptoms in gastric carcinoma is relatively short: in 62.6 per cent of cases under one year and in 81.3 per cent under two years. Sixty per cent of the cases occur between the ages of fifty and sixty-nine. The most common symptom is loss of weight. Therefore, weight loss with a short history in the age group mentioned should not be regarded lightly even in the absence of positive roentgen findings. Pain and vomiting, while of common occur-

rence, indicate advanced disease. The symptoms are governed largely by the location of the tumor.

The diagnosis of early cases, the differentiation between benign and malignant disease, and the identification of pathological types are often difficult or impossible by x-ray. Roentgenography is only one of the methods of examination and should be interpreted in the light of the clinical picture. Several illustrative cases are given.

II. Rodgers, discussing gastroscopy, states that this procedure should logically follow the x-ray examination. Often the roentgenogram will completely define the tumor, and in such cases gastroscopy will add nothing and may be dangerous. The roentgen examination should reveal information about the esophagus of value to the gastroscopist. Gastroscopy is indicated when the x-ray has demonstrated no lesion or one whose nature is uncertain.

Three types of cancer are recognized by the gastroscopist. The first type, an ingrowing polypoid type, presents a typical appearance and is usually readily recognized by x-ray. The second type, ulcerating carcinoma, is frequently identifiable as malignant, but not always. If an ulcerating lesion is found on or near the greater curvature it should be regarded as carcinomatous. When it is on or near the lesser curvature a fourteen-day trial of medical treatment is justified, followed by re-examination by gastroscopy. The third type, submucous infiltration, typically seen in leather-bottle stomach, is usually readily identified, but occasionally resembles benign atrophic gastritis. In these instances a re-examination should be made after two or three weeks of treatment.

Some information regarding prognosis may be obtained from gastroscopy, but this is not altogether reliable, for distant metastases may occur from the smallest lesions.

III. Stewart discusses chiefly the question of ulcer-cancer. Usually the malignant character of a carcinomatous ulcer can be readily identified pathologically. However, the appearance at the edges of some ulcers may be misleading. In some of these cases the clinical history may suggest the correct diagnosis.

The prognosis cannot be accurately determined from the type of carcinoma. Scirrhus carcinomas, however, usually have the worst outlook. This may be due to the fact that the diagnosis is made later than in other types.

IV. The outstanding fact affecting the surgical aspect of gastric carcinoma, as outlined by Taylor, is that the disease may be symptomless for a long period. The classical symptoms of pain, tumor, vomiting, and bleeding are of late occurrence. Weight loss and anorexia in middle-aged patients should arouse the diagnostician's suspicions. It may be a fatal mistake to believe that, if these early symptoms disappear under medical treatment, cancer is not present.

Only two methods of examination are of real value in the diagnosis, radiology and gastroscopy. Achlorhydria and occult blood in the stools may be suggestive, but are unreliable. Cancer may be present with a high acid and no bleeding. It might be supposed that examination of the stomach after laparotomy would be the best way to make a diagnosis. Sometimes, however, even with the stomach opened, the surgeon may have great difficulty in finding and identifying a small lesion.

Laparotomy is the only way to determine operability

in most lesions, certainly in early ones. In the presence of metastasis, the local growth is the least important factor in the prognosis, so there is little point in removing it. If there is, or is apt to be, obstruction at the pylorus, gastro-enterostomy is the best method of treatment.

In the absence of visible metastasis the operability of the growth becomes a subjective attribute of the surgeon, depending on his skill, experience, and willingness to take a risk in the hope of cure. For this reason, statistics from individual surgeons may be misleading. In the author's opinion every risk should be taken, with removal of all that seems necessary, including the whole stomach, its omentum, the spleen, the transverse colon, and the tail or body of the pancreas if necessary to accomplish cure. Of 38 consecutive patients undergoing radical operation for gastric carcinoma, 17 had one or more contiguous viscera removed; 7 died as a result of the operation but 10 were alive and well at the time of this report, from one to four years. Among the 21 cases in which such radical surgery was not done, there were only 2 operative deaths, 8 died from recurrence, and 11 were alive and well, after one to four years.

About one-sixth of the gastric cancers are in the cardia. Here diagnosis by x-ray and gastroscopy is most difficult. Successful surgical removal in a late stage is also very difficult. It would seem, then, that if a lesion in the cardia is suspected, early laparotomy is justified.

V. Fairchild and Shorter treated 15 cases directly with x-ray at operation. In only 8 of these was the disease thought to be localized enough for treatment to be effective. Of the 8 patients, 1 lived two years, 2 lived fifteen months, 1 seven months, 1 four months, and 1 three months after treatment. While the series is small and the results not too good, the authors feel that the method offers encouragement.

VI. Jennings points out that statistical studies show the incidence of gastric cancer to be about the same in widely different environments. There is not sufficient information for adequate evaluation and comparison of different methods of treatment. Survival rates after radical operations compare favorably with those after similar operations for other malignant disease.

SYDNEY J. HAWLEY, M.D.

Roentgen Findings in Primary Duodenal and Paraduodenal Malignant Lesions. J. R. Kline and G. J. Culver. *Am. J. Roentgenol.* 58: 425-438, October 1947.

Cancer of the duodenum has long been a diagnostic problem. Before the advent of metastases or involvement of adjacent structures, it is very difficult to make the correct diagnosis by clinical means, and in only a few instances has the condition been recognized preoperatively by roentgen examination. Probably the most constant finding is occult blood in the stool; an accompanying secondary anemia may be present. Upper abdominal pain is the most common complaint.

In the *suprapapillary* portion of the duodenum, the lesion is usually a constricting scirrhous adenocarcinoma, narrowing the lumen and infiltrating and eroding the mucosa. On fluoroscopy, a very small protuberance jutting into the lumen, with destruction of the mucosa over this area, may be seen. There is no associated spasm or tenderness such as is found in inflammatory lesions of this region.

In the *peri-ampullary* portion, the lesion may be a

constricting adenocarcinoma, as described above, producing the same signs and symptoms. In addition, this lesion, if it arises near the papilla of Vater, may very early infiltrate the papilla, causing a blockage of the flow of bile into the duodenum. In this type of carcinoma of the duodenum, jaundice and light-colored stools may occur very early.

Papillary adenocarcinoma may also occur in the peri-ampullary portion of the duodenum. Early in the course, before the base has infiltrated the mucosa and submucosa enough to cause constriction of the lumen, there may be no symptoms referable to the lesion.

Carcinoma of the *infrapapillary* portion of the duodenum is less common than of the other two portions. The lesion, which usually occurs near the duodenojejunal junction, is most often an infiltrating, constricting adenocarcinoma. Here, as in the suprapapillary portion, the growth is more scirrhous than glandular in nature. Carcinoma of this portion has a tendency to metastasize earlier than lesions elsewhere in the duodenum and, because of the location, symptoms appear somewhat later.

One case of primary carcinoma of the duodenum, two of carcinoma of the ampulla of Vater, and one of carcinoma of the head of the pancreas, in which the lesion eroded the mucous membrane of the junction of the first and second portions of the duodenum, are reported.

Post Bulbar Ulcer of the Duodenum. Robert P. Ball, Allan L. Segal, and Ross Golden. *Am. J. Roentgenol.* 59: 90-99, January 1948.

When a normal duodenal cap is seen in cases of gastrointestinal hemorrhage and in patients with peptic symptoms plus back pain, post-bulbar ulcer should be suspected and carefully sought.

The crater is not always seen in post-bulbar ulcer but there are eccentric narrowing of the lumen and an indentation of the wall at the level of the lesion. The authors have found that the lesion is best demonstrated in an exaggerated oblique view made with the patient on the right side in a horizontal posture, rotated slightly toward a prone or supine position, or with the head of the table lowered slightly.

The incidence of post-bulbar ulcer varies from 5 to 20 per cent as reported in various necropsy series of duodenal ulcer. This wide variation is thought to be due to some discrepancy in classification of cases near the apex of the bulb and possibly, in part, to a difference in the diligence of the search. ZAC F. ENDRESS, M.D.

Functional Intestinal Obstruction on a Congenital Neurogenic Basis in Infancy. Wolf W. Zuelzer and James L. Wilson. *Am. J. Dis. Child.* 75: 40-64, January 1948.

The study is based on 11 cases presenting a clinical picture of obstructive ileus in the absence of mechanical causes of intestinal obstruction demonstrable at operation or at autopsy. All the cases are reported in detail. The authors state that no previous attempt has been made to describe the condition as a clinical entity. The onset is in the first few days or weeks of life and evidence of a familial tendency has been observed. Abdominal distention, vomiting, and severe constipation are characteristic features. Physical examination demonstrates distention and tympany, audible and sometimes visible peristalsis. Later, the picture of paralytic ileus develops. There may be hard, dry impacted fecal

masses in the rectum, extremely difficult to evacuate. Paradoxical diarrhea at times appears.

The roentgenologic picture is virtually indistinguishable from that of ordinary intestinal obstruction. Survey abdominal films demonstrate gaseous distention and fluid levels. Usually the films suggest obstruction in the terminal ileum or in some portion of the colon. Barium studies have been of little aid and, because of the protracted retention of dried out barium, add to the difficulty and danger.

Enterostomy or colostomy was found to be the only method of relieving the condition. At operation no evidence of mechanical obstruction was found. In the more chronic, less severe cases a clinical picture somewhat resembling megacolon may gradually develop.

Histopathologic examination of the intestine in the cases reported revealed a consistent absence of the myenteric plexuses in the segments immediately distal to the obstructed gut. A congenital absence of the plexus is proposed as the cause of the condition.

ALTON S. HANSEN, M.D.

On Spontaneous Chronic Intestinal Invagination in Adults. P. Bjerre Hansen. *Acta radiol.* 28: 115-128, April 30, 1947.

After a brief review of the development of the roentgen diagnosis of intestinal invagination, with particular regard to chronic cases in adults, the roentgen signs of this condition are discussed. The author gives barium perorally in addition to, and combined with, the more common contrast enema and believes that this procedure has decided advantages. The proportion of cases of spontaneous invagination occurring in adults is variously given, from the 10 per cent of most authors to about 50 per cent in one Finnish series (Gullichsen; *Acta chir. Scandinav.*, Supp. 35, 1935). The etiology of invagination is discussed and it is pointed out that spontaneous cases can scarcely be very rare, either in adults or children.

The author reports 2 cases of intussusception in men of 29 and 35 years, both of which were considered spontaneous, although in one case appendicitis may have played a part. Both cases were characterized by acute onset with intense but indefinite, diffuse abdominal pain; this phase passed on to a chronic condition, with periodic abdominal pain and diarrhea, with liquid, mucous, non-hemorrhagic stools and considerable emaciation. Roentgen examination, with barium administered perorally and by enema, revealed ileocecal invagination in both patients. In one case complete disinvagination was achieved with the barium enema; in the second only partial reposition took place and operation was necessary. In neither case was a cause for the intussusception demonstrated roentgenologically.

Analysis of Roentgen-Ray Diagnosis in Carcinoma of the Cecum and Ascending Colon. Charles H. Brown and James R. Colvert. *Ann. Int. Med.* 27: 936-943, December 1947.

The purpose of this paper is to evaluate the barium enema findings in carcinoma of the cecum and ascending colon. The peculiar anatomy and the character of the pathological changes of this part of the digestive tract are responsible for certain difficulties in diagnosis. First, since the cecum is most distant from the anus, the cleansing enema is less apt to be effective there than in the rest of the lower bowel. Second, because of the

larger diameter of the cecum and ascending colon, obstruction is not usually produced as early as it would be elsewhere in the large bowel. Third, in view of the size of the cecum and ascending colon, it is particularly easy to overlook a filling defect, especially on the posterior wall. Fourth, carcinomas in this location are apt to be polypoid, producing late symptoms, signs, and roentgen-ray findings.

In a series of 50 cases of carcinoma of the cecum and ascending colon the diagnosis was made on the first barium enema study in 36 cases. Examination was repeated once or twice in 10 cases, either because the first examination was unsatisfactory, owing to poor preparation of the patient, or because the clinical findings were so suggestive of a malignant lesion that the clinician was not satisfied with the negative roentgen-ray report; in all 10 cases the malignant lesion was eventually demonstrated. Thus the roentgen examination was diagnostic in 46 out of 50 cases.

A filling defect was demonstrated in 32 of the cases and was the most common roentgen finding. Irregularity was present in 24 cases. In 8 there was a narrowing of the lumen, in 7 obstruction to the barium, and in 6 cases small bowel obstruction. One patient showed evidence of intussusception.

Two cases were difficult to diagnose due to the ptotic redundant position of the cecum, making palpation difficult. It is important that overfilling of the colon and ileum be avoided since loops of the ileum may then obscure the cecum.

STEPHEN N. TAGER, M.D.

Carcinoma of the Colon Secondary to Chronic Ulcerative Colitis. Thomas M. Johnson and Thomas G. Orr. *Am. J. Digest. Dis.* 15: 21-23, January 1948.

There is a considerable difference of opinion as to whether or not patients suffering from ulcerative colitis show an increased incidence of carcinoma of the colon. Those who believe so say that one can trace a case through the stages of inflammation, gland cell hypertrophy and hyperplasia, to adenoma and adenocarcinoma. Others are just as positive, after reviewing pathologic sections from cases of ulcerative colitis, that the pseudopolypoid tumors that occur never become true polypi, and that they may even regress and disappear. True polypoid tumors are apparently not caused by chronic infection. They never disappear unless the tumor happens to break away from its pedicle, and they often become malignant.

There is evidence that carcinoma of the colon develops more often in children who have ulcerative colitis and also that carcinoma occurs at an earlier age in patients with ulcerative colitis than in those not so affected. In a group of 40 cases from the literature, in which both conditions occurred, the average age at which carcinoma was discovered was forty years; the average age in patients without colitis is fifty-three.

The authors present two cases. One patient, a man aged 43, had an ileostomy performed for ulcerative colitis of the distal third of the transverse colon, the descending colon, sigmoid, and rectum. Three years later an advanced carcinoma of the cecum was found. [It should be noted, however, that there was no mention of colitis of the right side of the bowel.]

The second patient was a 32-year-old white man who had ulcerative colitis for sixteen years and was found at operation to have two carcinomas, one of which arose from a polyp.

JOSEPH T. DANZER, M.D.

Role of the Roentgenologist in the Diagnosis of Polypoid Disease of the Colon. Paul C. Swenson and Russell Wigh. *Am. J. Roentgenol.* 59: 108-121, January 1948.

The various forms of polypoid disease of the colon, namely, the single lesion, the limited multiple type, true multiple polyposis, and the polypoid manifestation of ulcerative colitis are discussed and illustrated.

Obscure gastro-intestinal bleeding calls for careful and repeated search of the colon to rule out polypoid lesions. If the pedicle or dimpling of the lumen at the site of attachment can be demonstrated, surgery is indicated without a second examination. In many of these, malignant change is already present at the base and excision is indicated, with a portion of bowel wall, if possible. When one polypoid lesion is found, the rest of the colon must be carefully examined because frequently others are present. Compression spot filming is very helpful in demonstrating them.

When dimpling or the pedicle cannot be seen, a subsequent examination is indicated to rule out a fecal mass before surgery is undertaken.

The various methods of examination—single- and double-contrast and compression techniques—and indications for each are discussed, along with the responsibilities of the roentgenologist to both patients and colleagues. Examples of all lesions mentioned are well reproduced.

ZAC F. ENDRESS, M.D.

Obstruction Due to Volvulus of the Sigmoid Colon. R. K. Gilchrist. *Arch. Surg.* 56: 79-91, January 1948.

Older patients with volvulus of the sigmoid colon usually give a history of repeated attacks of partial obstruction or of constipation, while younger patients often have a sudden onset of acute obstruction without an antecedent history. The patient is less sick than is usual in carcinoma, and the hemoglobin and red cell count are not much below normal. Although there is marked abdominal distention, there may be little vomiting. Pain is felt over the twisted loop and is more severe if the circulation is obstructed. The roentgenograms show remarkable gaseous distention, with loss of haustral markings. The dilated sigmoid loop usually reaches as high as the left leaf of the diaphragm, and the bowel wall at the flexion creases appears very thick. Barium enema examination occasionally demonstrates the markings of the twisted area.

Four cases, two showing the twisted area on the roentgenograms, are discussed, and a fifth case with volvulus of the cecum is included for contrast; this showed early associated small bowel obstruction, dehydration, failure of relief following intubation with a Miller-Abbott tube, and less characteristic x-ray findings.

The author advises resection of the redundant sigmoid loop, because of the high incidence of recurrence following conservative treatment. Cecal volvulus may be treated by untwisting and fixing the cecum to the pelvic wall, although some cases may require resection.

LEWIS G. JACOBS, M.D.

Extrinsic Lesions Affecting the Rectosigmoid. Richard H. Marshak. *Am. J. Roentgenol.* 58: 439-450, October 1947.

The roentgen characteristics of extrinsic lesions affecting the rectosigmoid are considered, especially as to their differential diagnosis from carcinoma of the

bowel. There are many pathological processes arising in the pelvis exclusive of the large bowel which, because of their proximity or secondary involvement, produce defects in the rectosigmoid. In some cases, the deformity of the rectosigmoid is typical, whereas, in others, differentiation from cancer of the bowel is impossible. The cases fall in the following groups: endometriosis; carcinoma of the cervix with a "frozen" pelvis; chronic inflammatory disease; ovarian carcinoma and ovarian cyst; effects of radiation therapy; uterine fibroid; sigmoiditis; lymphosarcoma and metastatic carcinoma; retroperitoneal tumors; and postoperative adhesions. The roentgen findings in these groups are discussed.

To simplify differential diagnosis, the cases have been divided into two main groups, according to whether the extrinsic pressure is due to inflammatory reaction or tumor (benign or malignant). Group I includes endometriosis, pelvic inflammatory disease, radiation injury to the bowel, and carcinoma of the cervix. The roentgen signs of the conditions in this group are fairly typical and differentiation from one another is usually not difficult. They can, however, be confused with the following intrinsic lesions involving the rectosigmoid: carcinoma, diverticulitis, lymphogranuloma venereum, and ulcerative colitis. Group II includes uterine fibroids, ovarian cysts, carcinoma of the ovary, and carcinoma metastatic to the pelvis. Some of the lesions in this group compress the rectosigmoid without being attached to the bowel wall and others attach themselves intimately to the serosa. Extrinsic unattached lesions are characterized by a gradual indentation which starts within the uninvolved area a short distance away from the actual edges of the compressing tumor and extends to the most protruding part of the tumor; the mucous membrane always remains intact. Attached extrinsic lesions produce sharply defined straight edges between uninvolved and involved bowel; in profile, only one side of the bowel is seen to be invaded.

Reproductions of a number of barium enema films illustrate some of the points brought out in the discussion.

Gastro-Intestinal Symptoms in Disease of the Urinary Tract: A Radiological Report. Milton Davis and Francis E. O'Neill. *Texas State J. Med.* 43: 567-569, January 1946.

This paper has to do with that group of patients who see their physicians for gastro-intestinal complaints, frequently vague, occasionally specific, which on careful examination are found to be the result of disease of the genito-urinary system. A distinct responsibility rests upon the radiologist to insist upon a thorough examination of the genito-urinary system in those cases which have been proved negative from a gastro-intestinal standpoint and in which the possibility of disease of the urinary system has not been completely eliminated.

Anatomically, there is a common sympathetic nerve innervation of the kidneys, upper third of the ureters, stomach, and upper gastro-intestinal tract. Whether or not this explains the relationship between the gastro-intestinal system and the genito-urinary tract is open to question, but it does offer a possible explanation of the nausea, vomiting, epigastric pain, and indigestion so frequently observed in pathologic conditions of the urinary system.

The diseases of the genito-urinary system most com-

moosly producing symptoms referable to the gastrointestinal tract involve the kidneys and upper third of the ureters. They include (1) renal tuberculosis, (2) congenital polycystic disease, (3) nephrolithiasis, (4) hydromephrosis, (5) renal ptosis, and (6) rarely, tumors of the kidney.

The author presents four illustrative cases. No illustrations accompany the article.

ERNEST S. KERESKES, M.D.

Study of the Gallbladder by Diagnostic Pneumoperitoneum. Antonio Lura and Axile Vivarelli. *Radiol. med. (Milan)* 33: 633-644, December 1947.

The authors have studied, by means of pneumoperitoneum, a number of patients suffering from occlusive jaundice. They believe that this means of examination permits determination of the volume of the gallbladder, its position in the abdomen, and its relationship to neighboring organs.

CESARE GIANTURCO, M.D.

Radiology of Biliary Ileus. Lidio G. Mosca and Bonfiglio A. Mateucci. *Prensa méd. argent.* 34: 2473-2481, Dec. 26, 1947.

The authors emphasize the necessity of systematic radiography in cases of acute abdominal crises. Numerous references are made to large clinics where radiologic exploration of the abdomen is practised in every abdominal emergency case. Opponents to this plan are also referred to, including one speaker at the German Congress of Surgery in 1928, who said "Here we do not photograph obstructions, we operate them." It has been argued that radiological exploration in an acute abdominal emergency entails a dangerous loss of time. Actually, the delay is usually to be laid at the door of the internist. The time which is lost between examination by the clinician and intervention by the surgeon may be considerably shortened by the early diagnosis made possible when radiology is employed. Frequently several days elapse during which various treatments—not always harmless—are tried, so that the surgeon is late in being called.

In view of the delays which are everywhere seen in bringing the "acute abdomen" to surgery, it is curious that objection is raised to the delay of one hour used in the employment of a simple radiological exploration, which often alone is sufficient for the correct diagnosis. Only with the aid of radiology can intervention be accurately planned and exercised in ample time, and with a minimum of trauma.

The authors refer to a triad of radiological signs typical of biliary ileus: (1) direct visualization of an opaque stone shadow in the lumen of the intestine; (2) dilatation of jejunal loops, producing a shadow simulating piled up coins (herring-bone appearance); (3) demonstration by ingestion of opaque substance of a cholecystoduodenal fistula. [The authors apparently are not aware of the value of gas in the biliary tract to demonstrate such fistulae.—J. T. C.] Demonstration of the stone itself in the intestinal lumen is exceptional. Gallstones in the bowel may appear in the form of an opaque ring or rings, or sometimes as homogeneous opacities; at other times as a central opaque nucleus surrounded by a density diminishing toward the periphery. They may present transparent zones, and in still other instances their opacity may be comparable to the soft parts.

Four Argentinian cases previously reported are cited and a fifth case is added.

JAMES T. CASE, M.D.

Chronic Recurrent Pancreatitis. A Clinical Study of Twenty Cases. Samuel N. Maimon, Joseph B. Kirsner, and Walter Lincoln Palmer. *Arch. Int. Med.* 81: 56-72, January 1948.

The purpose of this paper is to review the significant manifestations of chronic pancreatitis as observed in 20 patients and to direct attention to certain features which may be helpful in the diagnosis. Twelve of the patients were men. One-half the group were under fifty years of age. Recurrent bouts of severe pain in the upper abdomen constituted the chief complaint in 85 per cent. Symptoms had been present for less than one year in 40 per cent; in the remaining 60 per cent the average duration was four years. Physical findings were infinite and non-contributory. Jaundice of varying degree was present in 25 per cent of the patients. The oral test for glucose tolerance, carried out in 9 patients, yielded a diabetic type of curve in 8 and a flat curve in 1.

There was no constant relationship between the roentgen visualization of the gallbladder and the presence of pancreatitis. Non-visualization after oral administration of dye occurred in 5 cases. In 3 of the 4 patients operated on, in whom there was non-visualization of the gallbladder, no primary cholecystic disease was found. Cholelithiasis was noted at operation in 2 of 5 other patients in whom normal cholecystograms were obtained.

Abnormalities were found in 3 of 13 patients who had roentgen examinations of the stomach and duodenum. In one there was displacement of the lesser curvature by a mass posterior to the stomach (pancreatic cyst). In a second, dilatation of the stomach and first portion of the duodenum, demonstrated roentgenologically, was found to be due to narrowing of the second portion of the duodenum by an inflammatory mass involving the uncinate process of the pancreas. Dilatation of the second portion of the duodenum, in the remaining case, was caused by an inflammatory process which narrowed the third portion.

Pancreatic calcifications were demonstrated roentgenologically in 7 cases, localized in various parts of the pancreas in 4, and involving the organ diffusely in 3. There was no uniform pattern in the arrangement of the calcification, and the differentiation between calculi in the ducts and parenchymal calcifications could not be made clinically.

In 10 patients of the group (50 per cent), mild to significant disturbances in carbohydrate metabolism were found; in 3 the curves for glucose tolerance were elevated. Pancreatic calcifications were present in 4 of 7 patients with diabetes. Steatorrhea was present in 5 cases; pancreatic calcifications were demonstrated in 4 of these, and a diabetic state was present in all 5.

Surgical exploration was performed in 16 of the 20 patients, and various operative procedures were carried out. Seven patients were relieved of symptoms, 3 improved, 3 died; and in 3 there was no change.

Half of the patients in the series had had previous surgical treatment, without relief of abdominal pain. The possibility of pancreatitis must be especially considered in patients who continue to have attacks of pain simulating that of biliary colic after cholecystectomy has been performed.

Pancreatic Calculi. E. L. Eliason and Robert F. Welty. *Ann. Surg.* 127: 150-157, January 1948.

The reports of the last fifteen to twenty years indicate that pancreatic calculi are much more common

than is generally appreciated. The etiology is not clear. Most patients show no associated biliary tract disease. The authors consider that the primary factor is an alteration in the composition of the external secretion of the pancreas, with stasis perhaps playing a secondary role. Attacks of pancreatitis may give rise to areas of calcification but are not the source of most calculi.

Pancreatic calculi resemble closely those seen in the salivary glands and ducts. They are grayish white, hard, rough, and usually multiple, varying in size from a grain of sand up to 2.4 inches in diameter. They occur most commonly in the head of the pancreas and with decreasing frequency toward the tail. The disease predominates in the male and in the fourth and fifth decades.

The most common symptom is epigastric pain. Other findings are weight loss, nausea, vomiting, diarrhea, and jaundice. Diabetes occurred in 50 per cent of 66 cases collected from the literature.

The most important diagnostic measure is roentgen examination. This should include preliminary films, both anteroposterior and lateral or oblique. Many cases have been missed because a barium meal has been given before any films were taken, with the result that the overlying stomach and intestinal shadows obliterated those of the calculi. Being almost pure calcium, pancreatic stones cast a denser shadow than most biliary tract calculi. They may, however, be confused with these and with renal stones, calcified lymph nodes, and calcific plaques in the splenic vessels or aorta. Cholecystography and pyelography should aid in establishing the location of the calcifications with relation to the biliary and urinary tracts.

Surgery is the treatment of choice in severe cases. Procedures include removal of stones and/or partial pancreatectomy. Milder cases are treated conservatively. The authors' surgical approach is described.

Brief summaries of 9 explored cases are presented. The bibliography is extensive.

EDWARD E. LEVINE, M.D.

THE SPLEEN

Primary Splenic Abscess. Michael Gelfand. *Lancet* 2: 904-905, Dec. 20, 1947.

A case of primary splenic abscess in a young African adult is presented. Radiologically, there are three points which make the diagnosis of primary splenic abscess almost certain: (1) elevation of the left dome of the diaphragm, (2) contralateral displacement of the heart, (3) a gas shadow below the dome and above a distinct fluid level, though such a gas shadow may be mistaken for a hydropneumothorax or an eventration of the left dome of the diaphragm. After positive radiological proof has been obtained, the diagnosis is confirmed by aspiration. In the case reported here, 14 pints of watery chocolate-colored pus were evacuated from the abscess. A month later there was still a fairly profuse discharge from the abdominal sinus. The roentgenogram is reproduced.

THE MUSCULOSKELETAL SYSTEM

Ankylosing Spondylitis. William Lennon and Isabella S. Chalmers. *Lancet* 1: 12-15, Jan. 3, 1948.

The authors believe that ankylosing spondylitis and rheumatoid arthritis are one and the same disease, the

only difference being in the pattern and order of the joints affected. Rheumatoid arthritis gives essentially a centripetal distribution, and the hips and spine are often spared, even in severe cases. Ankylosing spondylitis, for some unknown reason, has a centrifugal origin and order of spread.

Of the 32 cases of ankylosing spondylitis observed in the Rheumatic Unit, Royal Free Hospital, London, only a small proportion had been accurately diagnosed before admission, though the recognition of the condition, even in its early stages, is not difficult. In this group, 12 patients first complained of low backache. In the early stages, only one patient spoke of stiffness without pain. There may be practically no clinical findings. Radiologic changes in the sacroiliac joints are almost always present by the time the patient seeks advice and it is these that establish the diagnosis. They consist of blurring and gradual obliteration of outline, perhaps at first unilateral but soon becoming bilateral. The erythrocyte sedimentation rate is usually raised in early cases, helping to exclude such diagnoses as fibrositis, intercostal neuralgia, lumbago, and neurosis. Tuberculous sacroileitis may be confused with ankylosing spondylitis but is almost always unilateral and its symptoms are non-remittent. Osteoarthritis of the spine can produce similar symptoms, but the roentgenograms clinch the diagnosis. Adolescent kyphosis must be considered. The girdle sensations may suggest tabes dorsalis, syphilitic pachymeningitis, or intra-abdominal and intrathoracic conditions. The rigidity of an advanced case may, at a glance, suggest paralysis agitans. Other causes of chronic backache (prolapsed disk, neoplasm, osteomyelitis, spondylolisthesis, etc.) should cause little difficulty in the differential diagnosis.

A regime such as is used at sanatoria for tuberculosis appears the ideal for patients with ankylosing spondylitis, and on general lines the treatment should approximate this as much as possible. Generalized ultraviolet irradiation had been given to all the cases in the series reported. The details of x-ray therapy in the treatment of ankylosing spondylitis are not discussed here. All the patients received this treatment but it is too early to decide on its efficacy.

Fibrous Dysplasia of Bone. Franklin B. Bogart and Allison E. Imler. *Am. J. Roentgenol.* 58: 478-484, October 1947.

Four cases of proved fibrous dysplasia of bone encountered on the radiologic service of a general army hospital in a three-year period are presented. A number of other cases were seen in which the roentgenograms suggested a diagnosis of fibrous dysplasia but in which a biopsy was not obtained. All of the cases reported here were relatively early, with changes confined to the skeleton. The bone changes as seen on the roentgenogram are suggestive and in some cases are diagnostic. The final diagnosis, however, is best made after careful consideration of the roentgen and clinical findings and correlation of these findings with the biopsy studies.

The authors mention particularly the differentiation of fibrous dysplasia from hyperparathyroidism and skeletal enchondromatosis (Ollier's disease). Hyperparathyroidism can usually be excluded by careful consideration of the bone changes and the distribution of the lesions if they are multiple. In case of doubt, studies of the blood chemistry should clarify the diagnosis. In differentiating fibrous dysplasia from skeletal en-

chondromatosis it is helpful to remember that fibrous dysplasia usually develops in childhood and not in infancy, usually has no gross lesions in the metacarpals, metatarsals, or phalanges, and has less shortening of the affected long bones.

In the discussion of this paper Dr. Aubrey O. Hampton states that he considers the diagnosis of fibrous dysplasia one of the easiest in radiology. The characteristic finding is a cystic-appearing lesion which is usually expansile, but the center, instead of being radiolucent, is more dense than could possibly be expected from a fluid-filled cyst. This is due to millions of partially calcified microscopic bone spicules in a matrix of fibrous tissue, producing the same effect "as if all bone architecture had been smudged by a thumb." The margins of the lesions are increased in density but do not have sharp peripheries except at the cortical margins, and even there the cortex may be destroyed; they fade off into the adjacent bone in a streaming fashion which has been compared to the appearance of a candle flame. This feature allows differentiation from enchondroma, which may otherwise closely simulate fibrous dysplasia.

Osteoporosis. Fuller Albright. *Ann. Int. Med.* 27: 861-882, December 1947.

Osteoporosis is characterized by a decrease in bone mass due to a lessened production of osteoid by the osteoblasts, such as occurs with advancing age, together with a continued normal degree of bone destruction. It is to be differentiated from two other conditions associated with too little calcified bone mass: osteomalacia, where there is a disturbance of the calcification of the osteoid, and osteitis fibrosa generalisata, where there is increased bone destruction.

Three factors entering into the decreased activity of the osteoblasts are discussed: (a) steroid hormones; (b) mechanical stresses and strains; (c) nitrogenous building blocks.

In respect to bone matrix, the steroids can be divided into anabolic (estrogens and androgens), anti-anabolic (adrenocortical "sugar" or "S" hormone), and anabolically inert steroids (progesterone). The age of entrance of the adrenocortical "N" ("nitrogen") hormone on the scene is termed the "adrenarche"; the age of exit the "adrenopause." The "adrenarche" is usually synchronous with the menarche; the "adrenopause" is normally considerably later than the menopause. The adrenal cortical "S" hormone is produced at the same level during childhood, sexual maturity, and senility. The osteoporosis of old age is partly to be attributed to loss of the gonadal hormones and of the adrenocortical "N" hormone and responds to estrogen and testosterone therapy. The osteoporosis of the post-menopausal state is to be attributed to the decrease in estrogen production and also responds to estrin and testosterone. The osteoporosis of Cushing's syndrome and of the adaptation syndrome of Selye is to be attributed to an excess of the anti-anabolic adrenocortical "S" hormone and responds to testosterone therapy. The adrenocorticotrophic hormone of the pituitary, by releasing "S" hormone from the adrenal cortex, likewise produces osteoporosis. Evidence is presented suggesting that the effect of testosterone in stimulating anabolism may be partly, but probably not wholly, due to its property of causing decreased production of the adrenocortical anti-anabolic "S" hormone rather than entirely to a direct anabolic property of its own.

The stress and strain factor is necessary for the stimu-

lation of osteoblastic activity. With the skeleton at rest, as during plaster immobilization, or with disuse from other causes, activity is halted and osteoporosis ensues.

As to the role of nitrogenous building blocks, evidence is presented indicating that serum albumin is an important precursor of bone matrix.

Calcinosis. A Brief Review of the Literature and Report of Two Cases. Frank T. Moran. *South. M. J.* 40: 840-844, October 1947.

Calcinosis is an infrequent affliction. The patients may show (1) localized calcific deposits around the joints, frequently bilaterally, in the skin, and in the subcutaneous tissues, or (2) diffusely in the skin, subcutaneous tissues, muscles, and tendons. The first type has been referred to as calcinosis circumscripta and the second as calcinosis universalis. A review of the literature yielded over a hundred case reports. Calcinosis universalis is found to occur mainly in the first two decades of life and about equally in the sexes. Calcinosis circumscripta occurs predominantly in females (about 85 per cent) and is rarely encountered before the second decade. In several cases familial tendencies have been reported.

The etiology of the disease is obscure. Some have attributed it to a derangement of calcium metabolism. In many instances the condition developed silently with no evidence of a precipitating factor. Scleroderma, sclerodactylia, and Raynaud's disease have been associated with calcinosis, particularly with the circumscripta variety. Forty per cent of cases of the latter type show scleroderma, as compared to 32 per cent of the cases of calcinosis universalis. Vasomotor spasm and also parathyroid gland disturbances have been suggested as playing a role in the pathogenesis of the disease but studies of calcium and phosphorus metabolism have not borne this out. When analyzed chemically, the deposits consist mainly of calcium phosphate and calcium carbonate. Sections of tissues from the areas of deposition show no evidence of necrosis or degenerative change.

Two cases of calcinosis universalis are described, in children under ten years of age. Roentgenograms showing calcium deposition in the soft tissues of the extremities, pelvis, and the axillary area of one of the patients are reproduced.

WILLIS MANGES, M.D.

Osteogenesis Imperfecta Tarda—Lobstein's Disease. Murray Clarke. *M. J. Australia* 2: 722-723, Dec. 13, 1947.

The classical triad of multiple fractures, blue sclera, and deafness is well known as one of the best examples of hereditary transmission of a pathologic condition. A case report is given to call attention to some other features associated with this syndrome which are less familiar, notably, in this instance, excessive laxity of the ligaments. This was first noticed when the patient was about eight months of age, manifested by clicking sounds as she moved her legs and arms. She was late in standing and had difficulty in walking. The sclerotics were blue, but hearing was apparently normal. There was no history of brittle bones in the family, but the mother's sclerotics were blue and she and her mother and brothers were deaf.

Discussing the radiologic findings, the author mentions the extremely slender shafts and the dense cortex

of the long bones. Transverse lines of greater density are apparent in the roentgenogram which he reproduces, possibly representing periods of growth disturbance or perhaps incomplete compression fractures, as suggested by Brailsford (*Radiology of Bones and Joints*, 3d ed., 1944). The appearance of the skull is quite characteristic. The bones are thin and the bitemporal diameter is increased, resulting in what has been described by Brailsford as the "tam o'shanter" appearance. There are also palpable angular projections above the level of the zygoma, causing the upper parts of the ears to project laterally and to be unduly prominent; the broad, wide forehead and small face give a triangular appearance to the head. Deafness, while it may be due to relaxed ligaments in the middle ear, is most generally attributed to otosclerosis, the hereditary character of which is quite well established.

Von Lobstein in 1833 applied the name osteopsathyrosis idiopathica to the general group of idiopathic brittle bones. Subsequent writers have suggested various classifications to include the large number of transitional forms. There are two main types. In one, multiple fractures occur before or soon after birth and the mortality rate is high. In the other, relatively few fractures occur and these only in the period between three and thirteen years of age. The fragility of bone, blue sclerotics, and laxity of ligaments are, according to Voerhoeve, all manifestations of an underlying hereditary hypoplasia of the mesenchyme. There is a congenital inability to form osteoblasts in contrast to the excessive proliferation of osteoid tissue in rickets.

SYDNEY F. THOMAS, M.D.

Traumatic Cortical Osteophytosis (Cortical Osseous Changes Appearing on Some Diaphyses Subjected to Microtraumatism). F. Saegesser. Schweiz. med. Wochenschr. 77: 1302-1304, Dec. 13, 1947.

The author reports the appearance of biopsy specimens from two young men with thickening of the cortex of the shaft of the tibia, which he believes throws light on the complex called by various writers osteoid osteoma, cortical osteoid, tumor-like periostitis, etc. In each case there was a history of repeated minor traumata, followed by chronic, disabling pain and x-ray demonstration of cortical sclerosis of the bone. The biopsies showed slightly different stages of progress, but the cortical changes were on the basis of focal areas of necrosis with subperiosteal new-bone formation.

The author feels that the production of this lesion follows a distinct pattern, beginning with chronic slight traumata, such as are sustained in cadence marching, hiking with full pack, etc., which produce vasomotor disturbances in the bone with interruption of nutrition and later focal areas of necrosis in the deeper part of the cortex. Repair then takes place by the formation of osteophytic new bone, which is laid down beneath the periosteum. At some stages of this syndrome the bone is abnormally fragile, and fractures may be produced by insidious means. [These cases appear to conform to "fatigue fracture" rather than osteoid osteoma.—L. G. J.]

LEWIS G. JACOBS, M.D.

Case of Glomus Tumor with Primary Involvement of Bone. Raffaele Lattes and David C. Bull. Ann. Surg. 127: 187-191, January 1948.

A report of a glomus tumor completely encased in the terminal phalanx of a thumb is presented. Roent-

genograms showed honey-combed areas of decalcification of the distal phalanx of the thumb, suggesting cysts or enchondromata. At surgery, the bone was found almost completely replaced by a jelly-like material, with a paper-thin cortex. The lesion was curetted, and the defect filled with a bone graft. The histologic features were typical of glomus tumor—"epithelioid" cells arranged in perivascular cuffs around numerous endothelium-lined spaces.

The pathogenesis of glomus tumors arising where no normal glomera have ever been found is briefly discussed. This is believed to be the second case reported of a glomus tumor completely encased in bone.

EDWARD E. LEVINE, M.D.

Pseudo-arthritis of the Carpal Scaphoid as a Late Sequel of Trauma. Francisco Fernández Rozas. Prensa méd. argent. 34: 2482-2498, Dec. 26, 1947.

The tendency of the scaphoid to undergo pseudo-arthritis is due to two factors: circulatory (faulty blood supply) and mechanical (faulty immobilization). To avoid the aphorism of Destot that "scaphoid fractures rarely heal; pseudo-arthritis is the rule," one should practice early diagnosis and early immobilization. Pseudo-arthritis carries with it the complication of aseptic necrosis of the fragments, and arthritis and arthrosis involving the radionavicular articulation and even the whole carpus.

JAMES T. CASE, M.D.

Pseudocoxalgia: (Calvé-Legg-Perthes' Disease): Radiographic Changes Outside the Femoral Head. H. G. Korvin. Proc. Roy. Soc. Med. 40: 886-891, December 1947.

On the basis of 90 cases with 102 affected hips, the author describes radiographic changes outside the femoral head in terms of recognized pathology.

Increased distance between the medial pole of the femoral head and floor of the socket was described by Waldenström in 1937 as an early sign of pseudocoxalgia and was ascribed to a capsular effusion. Korvin states that capsular effusions in involved hips are minimal and insufficient to produce capsular tension. Arthrograms conclusively reveal a translucent zone, indicative of soft tissue interposed between the femoral head and floor of the socket. In addition, excavation of the floor of the socket is evident whenever the Waldenström sign appears. Whitman has described a swollen, congested ligamentum teres upon opening a hip in a case of Perthes' disease and Korvin is of the opinion that the Waldenström sign is due to a swollen, hyperemic, pulsating ligamentum teres which displaces the femoral head laterally and erodes the floor of the socket.

The author does not regard the "cyst-like" areas that appear in the acetabulum and femoral neck as extensions of the osteochondritic process from the capital epiphysis. Osteoporosis of the neck and socket, in the early stages, is diffuse and homogeneous. As it continues, atrophy becomes more marked in some places and "cyst-like" areas appear. When the margins of bone are affected, the appearance suggests actual defects. Similar cyst-like spaces are to be seen in other conditions, as hypertrophic and rheumatoid arthritis, osteomalacia, occasionally in Paget's disease, and most characteristically in parathyroid disease. They disappear when the underlying condition is cured. Deformities that persist are those acquired during and as a result of the severe bone atrophy.

The main factor that determines the shape of the upper end of the femur is the deformity acquired by the necrotic femoral epiphysis before it regenerates. This, however, is not the only factor. Deformity of the femoral neck may appear very early, before manifestations of disease become visible in the capital epiphysis. The change is produced by osteoporosis of the femoral neck which permits tilting of the femoral epiphysis, similar to that seen in epiphyseolysis, but differing from it in that the neck rather than the epiphyseal line is involved. This finding occurs in about 50 per cent of the cases and may appear before any changes are demonstrable in the femoral head.

LOUIS BERNSTEIN, M.D.

Evaluation of Treatment of Slipping of the Capital Femoral Epiphysis. Ignacio Ponseti and Chester K. Barta. *Surg., Gynec. & Obst.* 86: 87-97, January 1948.

The authors reviewed 73 cases of "slipped" femoral epiphysis seen at the University Hospital in Iowa City from 1932 to 1947. In 19 cases the condition was bilateral. This series of patients was divided into 5 groups, as follows:

(1) Preslipping stage: 5 patients, 1 with bilateral slipping. The authors believe that this potentially dangerous stage should be treated by drilling and pinning as soon as the diagnosis is established.

(2) Acute slipping or fracture separation of upper femoral epiphysis: 19 patients, 1 with bilateral acute slipping. Best results were obtained by early and gentle reduction of the fracture separation, maintaining the reduction by two or three threaded Steinmann pins. The authors advise that when these patients are seen more than two weeks after the accident, no attempts should be made to reduce the slipped epiphysis.

(3) Chronic slipping with minimal to moderate displacement: 31 patients, 4 with bilateral chronic slipping. In this series best results were obtained by rest from weight-bearing, plaster cast immobilization, or by drilling and pinning of the upper femoral epiphysis.

(4) Chronic slipping with marked displacement: 21 patients, 5 with bilateral chronic slipping. The prognosis in this type proved to be poor under any form of treatment. The authors state that treatment should be directed toward preventing further slipping and hastening fusion of the epiphysis.

(5) Residual deformities: 4 patients, 1 with old bilateral slipping.

Three cases are recorded in which marked narrowing of the joint space was demonstrable roentgenographically following slipping of the upper femoral epiphysis.

RICHARD C. RIPPLE, M.D.

Painful Heels in Children. E. S. R. Hughes. *Surg., Gynec. & Obst.* 86: 64-68, January 1948.

The author reviews the literature of painful heels, in which he found mention of fourteen roentgen signs of this entity. He gives a description of the normal variations in the appearance of the ossifying epiphysis of the os calcis and describes the clinical and roentgen signs of painful adolescent heels as observed in a series of 23 patients six to fourteen years of age (17 boys and 6 girls). In 16 cases the symptoms were bilateral. Pain was intermittent but persistent, often precipitated by exercise and referred to the sides, the back, or the inferior aspect of the heel. In most cases there was a complete absence of physical signs. Roentgenograms showed a varied appearance. Sometimes the epiphyses

appeared enlarged or flattened or irregular or segmented; sometimes mottled and fluffy. The epiphyseal line was often cloudy and united the epiphyses to the irregular posterior surface of the os calcis.

The cause of this picture is discussed. There is lack of any histologic proof of disease, and it is impossible to agree that the films reported as abnormal differ in any way from a normal series. A follow-up examination revealed no residual anatomical abnormality in the heel, clinically or roentgenologically, despite the fact that pain sometimes persisted after fusion of the epiphysis and diaphysis. It is concluded that there is no proof that osteochondrosis or epiphysitis is the underlying cause of adolescent painful heels.

JORGE DE LA FLOR, M.D.

GYNECOLOGY AND OBSTETRICS

Diagnosis of Ectopic Pregnancy by Hysterosalpingography. Børge Nielsen. *Acta radiol.* 28: 185-198, April 30, 1947.

By using perabrodil as a contrast medium the author finds the possibility for diagnosis of ectopic tubal pregnancy more certain than with iodized oil. In his series of 25 cases it was possible to establish the diagnosis in 11 and suspect it in 1; furthermore tubal pregnancy was ruled out in 12 of the remaining 13 cases and was doubtful in only 1 case.

The roentgenologic findings fall into four rather sharply defined groups. (1) The implantation is identified as a rounded filling defect in the tube, with a striped appearance over the surface. (2) There is a partial outline of the proximal portion of the implantation with a typical striped appearance over part of the surface. (3) Only the semilunar surface of the obstructing mass is visualized. (4) There is failure of filling of the tube on the side of the pregnancy.

Differential diagnosis includes the rare neoplasms and possibly tuberculous salpingitis with caseous degeneration. Of probably greater importance is the ability to rule out tubal pregnancy when the history is suggestive. The author feels that there is no particular risk involved if the examination is done properly.

ELIZABETH A. CLARK, M.D.

Diagnostic and Therapeutic Aspects of Kymographic Uterotubal Insufflation with Comparative Observations on Hysterosalpingography. I. C. Rubin. *J. Obst. & Gynaec. Brit. Emp.* 54: 733-745, December 1947.

In 1914, Cary and Rubin independently reported the injection of x-ray opaque substances into the uterus for the specific purpose of outlining the fallopian tubes. Some work had been done prior to 1914, but these early attempts were rather crude, the shadows obtained were poor, and evidently some reactions were produced. Lipiodol was widely used as a medium after its introduction into gynecologic practice by Heuser in 1923. More recently certain advantages have been claimed for "viscorayopaque" (see Norment: *Am. J. Obst. & Gynec.* 49: 253, 1945. *Abst. in Radiology* 45: 630, 1945), and this the author regards as the nearest approach to the ideal radiopaque substance yet attained.

Hysterosalpingography, however, especially with the iodized oils, has the disadvantage that the medium may be retained in stenosed or obstructed tubes, where it may induce further pathological change. In view of this fact, the author tried first oxygen and later

carbon dioxide to establish the fact of tubal patency or non-patency. Oxygen was found to cause shoulder pain lasting from one to several days, but carbon dioxide was not attended by this disadvantage. It leaves no residue and can be used repeatedly for therapeutic purposes. The technical procedure, employed for the past twenty-two years, includes the use of a kymograph in combination with a manometer and tank of carbon dioxide and is designated as kymographic uterotubal insufflation.

By carefully noting the radiation of the momentary pain which accompanies the insufflation, the site of obstruction can be determined, as has been proved by roentgenographic control. The author stresses one point particularly as favoring insufflation as compared with injection of the iodized oils, namely, the hazards of oil injection. In a collected series of lipiodol studies, infections were found in one of every 230 cases; in a series of insufflations gathered from various sources, the incidence of infection was only one in 1,705 cases. An analysis of two other series showed an incidence of complications of about one in 300 cases with lipiodol injection and 1 in 423 with insufflation.

In a questionnaire covering over 80,000 insufflations 15 embolisms were reported, with 3 deaths. It is significant, however, that embolism occurred only when air or oxygen was used. No deaths have been reported with the use of carbon dioxide. The tremendous solubility of carbon dioxide in equal volumes of fluid and its rapid resorbability probably account for this safety factor. Numerous criteria for the diagnosis of disease with kymography and tubal insufflation are given.

SYDNEY F. THOMAS, M.D.

THE GENITO-URINARY SYSTEM

Infantile Amaurotic Familial Idiocy and Congenital Hydronephrosis of a Solitary Kidney: Urethrocytography as a Diagnostic Aid. Bernard L. Lipman, Bernhard J. Statman, Robert A. Schless, Paul S. Friedman, and Leon Solis-Cohen. *Arch. Pediat.* 65: 25-29, January 1948.

The existence of congenital structural defects in combination with infantile amaurotic familial idiocy is uncommon. A case report is given demonstrating these features, as well as the value of urethrocytography in the diagnosis of congenital hydronephrosis.

The patient was a male infant of three months, with intermittent fever, increased blood urea nitrogen, and albuminuria. Subcutaneous urography was unsatisfactory on two occasions; on a third it indicated a possible left hydronephrosis. Urethrocytography was then done. The left urinary tract was well visualized, but the right was not demonstrable. Dilatation of the left ureter, renal pelvis, and calices was apparent. The conclusion was drawn that a solitary left kidney existed, with a congenital hydronephrosis, plus infection. Subsequently the diagnosis of amaurotic familial idiocy was made. An exploratory operation was performed, at which it was not possible to do much that was constructive.

[To the abstractor this history emphasizes the importance of retrograde examination when accurate information is required. Time is wasted and false conclusions may be drawn by the performance of so-called subcutaneous urography. Wesson, in his excellent text, states that he has never yet operated upon the evidence furnished by an intravenous pyelogram.]

PERCY J. DELANO, M.D.

Co-existing Hypernephroma and Tuberculosis. S. A. MacDorald. *Canad. M. A. J.* 57: 572-575, December 1947.

Only 21 microscopically proved cases of associated renal cancer and tuberculosis could be found in the literature. An additional example is reported.

A 52-year-old woman was admitted to the hospital because of several episodes of hematuria during the preceding eighteen months. She had rather severe renal colic on two occasions and had passed clots. There was a history of earlier surgical incisions in the region of the right anterior superior iliac spine for some suppurative process which occurred during adolescence, and pus was said to have drained from the wounds for a period of months. Since then the patient had had definite limitation of motion in the right hip joint. Physical examination showed clear lungs, moderately elevated blood pressure, tenderness in the right costovertebral angle, gross scoliosis of the spine, and ankylosis of the right hip. Cystoscopic examination revealed blood emerging from the right ureteral orifice. The bladder mucosa was injected and suggested tuberculous ulcerations. No acid-fast bacilli were found in the urine.

A plain film of the abdomen showed the right kidney shadow to be at a higher level than the left and the lower pole of the right kidney to be grossly enlarged. A bilateral retrograde pyelogram showed the left side to be normal. The right renal pelvis and calices were grossly distorted. The upper major calices were enlarged, and the minor calices were bluntly dilated. The middle caliceal areas failed to fill. The lower calices were only partly visualized, due to apparent compression. A second pyelogram showed expansion and distortion of the lower caliceal system on the right due apparently to an intrinsic renal tumor. The possibility of a combined inflammatory and malignant process in the same kidney was entertained preoperatively. X-ray films of the chest were negative for metastases but showed quiescent tuberculous foci in both apical regions.

A right nephrectomy was done and a circumscribed tumor was found measuring approximately 3.5 cm. in diameter, occupying most of the lower pole. The upper major calyx was broadened and shortened and there was blunting of the minor calices. Microscopic study showed the tumor to be a typical hypernephroma. Sections taken in the upper caliceal areas revealed a typical tuberculous reaction, with scarring, atrophy, and numerous discrete and conglomerate tubercles.

The author presents a discussion of a possible relationship between the two lesions. He raises the question of whether the patient did not have a tuberculous arthritis in youth in view of the hip findings. Some experimental work by Cherry in 1929 indicates that he increased the incidence of spontaneous cancer in mice by about 10 times through injections of small numbers of tubercle bacilli subcutaneously. Studies of several autopsy groups, however, reveal little effect of one disease upon the other and indicate that the combination is apparently a coincidence. WILLIS MANGES, M.D.

Diverticulum of the Male Urethra: Report of a Case. Harry J. Repman, Jr., and John W. Warren, Jr. *J. Urol.* 59: 44-46, January 1948.

In 1928 Lowsley and Gutierrez (*Rev. de med. y cir. Habana* 33: 229, 1928) found in the literature 108 cases of diverticulum of the male urethra and reported 6 new cases. A review of the English literature since

that time has yielded 31 cases, and to these another is added. It supports Fagerstrom's theory (J. Urol. 49: 357, 1943) that diverticula may occur following injury to the nerves supplying the urethral musculature, since all the clinical symptoms dated from a prostatectomy. A urethrogram made with skiodan, showing the large-mouthed diverticulum in the anterior urethra, is reproduced.

Heterotopic Bone Formation Following Suprapubic Prostatectomy. B. S. Abeshouse. J. Urol. 59: 50-66, January 1948.

Bone formation in the wound following abdominal operations is a rare condition and is more likely to occur in supraumbilical abdominal scars. The author presents a case of ossification occurring postoperatively in a suprapubic prostatectomy scar, bringing the total number of reports of such cases to 18. He discusses experimental heterotopic bone formation in transplantation experiments in animals, but points out that the etiology and pathogenesis are different in human extraskeletal heterotopic bone formation. In the latter, the most likely explanation of the new bone formation is the metaplasia of connective-tissue cells into bone cells, *i.e.*, young fibroblasts may become osteoblasts under such stimuli as (a) the infecting and irritating action of urine or pus in the operative area, (b) the irritating action of a suprapubic tube, (c) the availability of calcium and phosphorus salts in the urine to impregnate the affected tissue.

The heterotopic bone following suprapubic operation is found in three sites in the wound: (a) in the superficial scar, (b) in the deep fascia of the rectus sheath, (c) in young connective tissue around the suprapubic fistula. It is present as spicules or as large flat plates of bone and is true cancellous bone, histologically. The bony mass reaches its maximum size early (one or two months after operation) and remains stationary for a long period.

The diagnosis is established by palpation. A plain film may show an opaque shadow of bony density. Plain or stereoscopic films—anteroposterior, lateral, or lateral-oblique—will reveal the relation of the osseous

mass to the pubic bone. There may be no symptoms, but the spicular bone may delay closure of a suprapubic fistula. Radiography should therefore be employed in every case of persistent fistula following suprapubic operation to exclude this possibility. Occasionally dull pain and urinary disturbances due to pressure of the mass occur.

Heterotopic bone formation must be differentiated from (1) foreign body in the wound, (2) osteitis of the pubic bone, (3) hematoma of the rectus muscle, (4) neoplasms of the pubic bone, (5) muscle tumors, (6) interstitial syphilitic myositis, (7) pubic exostosis, (8) congenital osteoma cutis, and (9) calcification of a primary or metastatic tumor.

Treatment consists in surgical excision of the growth. There have been no recurrences reported.

The author has tabulated the findings in the reported cases and presents in detail his own case, in which the heterotopic bone occurred in the deep rectus fascial layer of the scar, six weeks after a suprapubic prostatectomy for benign prostatic hypertrophy. A bony mass $5 \times 3 \times 0.5$ cm. was excised.

D. B. NAGLE, M.D.

THE BLOOD VESSELS

On Venography of the Lower Extremities. J. Friemann-Dahl. Acta radiol. 28: 199-207, April 30, 1947.

The author has used venography as a method to study the alterations in venous circulation in the patient at bed rest. Thus depending on the amount of extension of the knee and upon the thickness of the calf, the pressure of the calf against a hard mattress produced slowing of flow of the medium through both the deep and superficial circulation to the point of occlusion if pressure were great. Studies made of both legs crossed gave similar findings. Motion increased emptying significantly, and studies made through a plaster cast showed no delay, possibly because of the uniform support afforded by the cast. Finally, it was shown that although a tight bandage was placed over the knees of a patient on the operating table, the rubber mattress prevented compression and only minimal changes were produced.

ELIZABETH A. CLARK, M.D.

RADIOTHERAPY

The Time Factor in Radiation Therapy. Charles L. Martin. Am. J. Roentgenol. 59: 1-8, January 1948.

The author reminds us of the well established but often forgotten fact that the time factor is an important one in measuring tumor doses. Curves reproduced show that the figure of 6,000 gamma roentgens, considered by Paterson and Parker to be cancerocidal when delivered over a period of 168 hours, gives only 3.2 threshold erythema doses when the time factor is considered (instead of 6 T.E.D.). Martin compares his own technic of fractionated 200 kv. therapy for large skin cancers in terms of T.E.D. and finds that his dose was 3.2 T.E.D., using the time factor.

A useful time-saving chart is given, showing values in T.E.D. delivered at various points around low-intensity radium needles in 168 hours. It was worked out by adding the time factor to Quimby's data on the distribution of radiation values around linear radium sources.

The paper concludes with some very practical ideas on the simplification of implantation patterns in tumors of various sizes and shapes. Thus by using the chart of dosages and sketching the implantation pattern to scale, it becomes an easy matter for anyone to plan the correct radium dosage for any tumor which can be treated in this manner.

ZAC F. ENDRESS, M.D.

Medical Progress. Radiation Therapy. Samuel A. Robins, Bernard E. LeVine, and George White. New England J. Med. 238: 55-60, Jan. 8, 1948.

A review of recent literature, with a comprehensive bibliography.

Preoperative Roentgen Treatment in Breast Cancer. Sigvard Kaas. Acta radiol. 28: 153-168, April 30, 1947.

The author analyzes the results obtained at the Radium Center in Copenhagen in breast cancer treated by

radical surgery either followed or preceded by roentgen therapy and compares them with the results obtained in various other hospitals. While he finds nothing in the literature which with certainty proves that preoperative roentgen irradiation improves the results of surgery, his own material, consisting of 301 patients, 137 of whom have been followed five years or more, seems to indicate that such may be the case. Nothing appears to be gained by greatly intensifying the preoperative roentgen dose; 1,000 to 1,500 r to each field is considered sufficient.

The frequency of distant metastases without simultaneous local recurrence was the same in the preoperatively and postoperatively irradiated groups, but local recurrence was much more frequent in the latter group. This may explain the better results obtained in the patients treated preoperatively. The incidence of local recurrence in the cicatrix and in the axilla and the frequency of metastases in the ipsilateral supraclavicular region would indicate that irradiation should be directed to fields over each of these sites.

Carcinoma of the Esophagus. An Analysis of 145 Cases with Special Reference to Metastases and Extensions. George J. Taquino and Gerald F. Joseph. *Ann. Otol., Rhin. & Laryng.* 56: 1041-1051, December 1947.

During the eleven-year period ending Dec. 31, 1946, 132 indubitable cases of carcinoma of the esophagus were observed at Charity Hospital of Louisiana at New Orleans. Another 13 cases were observed over the same period at the Hôtel Dieu Sisters' Hospital in the same city. At Hôtel Dieu, which receives private patients, exploration was done in one case, which was found to be inoperable. At Charity Hospital, which receives only indigent patients, 18 operations were done with the expectation of resection and 24 patients were treated with radiation. Seven patients were found to have inoperable growths. Of the 11 on whom resection was carried out, 8 did not survive the immediate postoperative period, and 1 died of recurrent disease in three months. The other two were lost from observation at the end of a year. Among the patients treated with radiation, 6 of 13 in whom cure was attempted were alive at the time of the report, from nine to eighteen months after treatment. The 2 surgical patients who may have survived and the 6 known to be living after irradiation represent the total salvage in these 145 cases. Seventy patients died under observation and in the rest death was not long postponed. This is the tragic clinical picture of a form of cancer which ranks fourth in males beyond the age of twenty and accounts for at least 8 per cent of all cancer deaths.

Regarding sex, age, race, and regional distribution of cases, the statistics from Charity Hospital do not vary greatly from other reported statistics. As in all other series, dysphagia, unexplained hoarseness, and hicoughs were the first presenting symptoms. In 124 of the 145 cases in this series the diagnosis of carcinoma of the esophagus was confirmed by autopsy or biopsy. The majority of the cases, as usual, were of the squamous-cell variety.

The esophagus is a particularly dangerous site for a malignant process. It is located in the midst of important and vital structures; it has no outer serosal covering, and it is richly supplied with lymphatics in the submucosal and muscular coats. As a result of the favoring circumstances, the growth tends to spread

longitudinally, to erode through the esophageal wall with consequent mediastinitis, and to reach mediastinal lymph nodes and other structures through the convenient lymphatic channels.

In 38 of 101 patients who survived the period of observation, metastases were identified by clinical methods in sixteen different sites, including six different groups of lymph nodes. In 30 of 44 patients submitted to autopsy, metastases were identified in nineteen different sites, including seven different groups of lymph nodes. The disparity between the sites of metastases identified clinically and at autopsy indicates the number probably overlooked in the group studied only by clinical methods. In 20 patients the disease had extended to adjacent organs. The sites of extension numbered twelve, and complications had occurred in 17 of the 20 cases. Altogether, metastases or extensions or metastases and extensions were identified in 88 patients. As a result, this group, which comprised 60.7 per cent of the total number, was automatically excluded from the benefits of surgery, and probably, in most instances, of radiation therapy as well.

Cachexia and inanition are the commonest causes of a fatal outcome, with pulmonary and mediastinal complications next in order.

Lesions of the intrathoracic portion of the esophagus are amenable to resection and re-establishment of continuity of the gastro-intestinal tract by bringing the stomach (or a loop of jejunum) into the thoracic cavity. Lesions of the extrathoracic portion can be treated by resection and re-establishment of gastro-intestinal continuity by bringing the stomach or a loop of jejunum through the dome of the thorax into the neck and anastomosing it to the stump of the esophagus.

The results of radiation therapy at the New Orleans Charity Hospital, recounted above, may be regarded as hopeful, though still not good enough to warrant optimism. The technic employed was that of Smithers and his associates (*Am. J. Roentgenol.* 49: 606, 1943. *Abst. in Radiology* 41: 613, 1943.), permitting the delivery of an average tumor dose to involved segments of the esophagus. It is particularly effective in lesions of the middle third, in which surgery is particularly difficult. The method is not without danger, however, carrying a risk of perforation into the great vessels or mediastinum.

STEPHEN N. TAGER, M.D.

Transvaginal High Voltage Therapy for the Treatment of Carcinoma of the Cervix. Charles A. Behney. *Surg. Clin. North America* 27: 1550-1554, December 1947.

For carcinoma of the cervix, the author uses 200-kv.p. therapy transvaginally, with cones 1.5 to 3.6 cm. in diameter, and a distance of 50 cm. from target to lesion. The cones are lined with 0.5 mm. lead and have either straight or beveled ends. Hard rubber obturators for introduction have been replaced by an inflated rubber prostatic balloon, which protrudes from the end of the cone and is easily removed by deflation after insertion is completed. This gives better protection to the soft tissues and permits better exposure of the lesion. The cone adaptor is equipped with a periscope for visualization of the lesion after attachment of the cone to the therapy machine.

A well padded flat-topped wooden table, 27 inches high with stirrups at one end, is used to place the patient on for treatment. An 18-inch leaf is hinged to the foot

of the table. By elevating this, the pelvis can be raised 6 to 12 inches, to bring the vaginal outlet in line with the adaptor. The patient is placed in the lithotomy position with her buttocks on the edge of the hinged leaf.

Prior to transvaginal therapy, external irradiation (200 kv., 50 cm. distance, 0.5 mm. Cu and 1.0 mm. Al filtration) is given over the pelvis, through four 15 × 20-cm. ports (two anterior and two posterior) for a total of 2,000 r (in air) over each field (3,000 tissue roentgens). If the external radiation is well tolerated, the transvaginal therapy is given coincidentally. If not, transvaginal therapy is postponed until external radiation is completed. The transvaginal dosage is 500 r on alternate days, to a total of 6,000 r in air (6000 tissue roentgens). A second series of external irradiation treatments similar to the first is given six to eight weeks after completion of the cone therapy.

In 33 Stage IV cases treated in this manner, the average life duration was twice that obtained in a series of 161 cases treated with external irradiation alone. In Stage III cases there was no apparent difference in duration of life as compared with external irradiation plus radium irradiation. The author, however, prefers this method to external irradiation and radium insertion as he believes there is less necrosis of normal tissue, very little distortion follows healing, and, if surgery is necessary, the technical difficulties of operation are reduced.

BERNARD S. KALAYJIAN, M.D.

Treatment of Cancer of the Bladder by Radium Needles. M. Lenz, G. F. Cahill, M. M. Melicow, and C. P. Donlan. *Am. J. Roentgenol.* 58: 486-491, October 1947.

Forty-four patients with histologically proved cancer of the urinary bladder in or near the trigone were treated by suprapubic cystostomy and implantation of radium needles. Twenty-five of these patients died within a year, 8 in two years, 1 in three years, all presumably of persisting or recurring cancer. One patient died fifteen years after treatment, of perinephritic abscess; no cancer was found at autopsy. Nine patients are living and appear free from clinical evidence of cancer from five to fifteen years after treatment. Results were best in small papillary tumors, though some of the larger infiltrating cancers responded well to treatment. A tumor dose of 8,000 gamma roentgens was found preferable to smaller doses.

Treatment of Polycythemia Vera by Spray Irradiation. Wyman Richardson and Laurence L. Robbins. *New England J. Med.* 238: 78-82, Jan. 15, 1948.

This is a discussion of a method of treating polycythemia vera by spray irradiation as practised at the Massachusetts General Hospital. Physical factors are: 200 kv., 0.5 mm. Cu plus 1.0 mm. Al filter, h.v.l. 0.92 Cu, 215-250 cm. distance, and a field covering the body from the shoulders to the knees. The dose is 20 to 30 r per day divided between the anterior and posterior field, with a total of 300 to 500 r for the series. The amount of treatment is determined by the white cell count, which is followed closely. Irradiation is discontinued when this count drops to 5,000 or 6,000. The red count will not be affected for one or two months after completion of the treatment.

The authors' series of patients numbers 28, of whom 12 have died—2 of pulmonary disease, 1 of carcinoma of the breast, 1 of uremia, 6 of arterial thromboses. Sixteen patients are living quite comfortably.

Irradiation is considered a treatment of choice in polycythemia vera and is believed to afford the best chance of a comfortable existence. Radiophosphorus has not been used and no comparison with this newer method of treatment can be made.

JOHN B. MCANENY, M.D.

Lymphosarcoma of the Tonsil in a Three-Year-Old Child. S. M. Morwitz. *Ann. Otol., Rhin. & Laryng.* 56: 892-897, December 1947.

A case of lymphosarcoma of the tonsil in a child of three years is reported, with rapidly fatal outcome. Although lymphosarcoma is known to be radiosensitive, roentgen therapy in this instance was ineffective. Autopsy revealed a generalized lymphosarcomatosis involving especially the cervical and mesenteric lymph nodes and the left kidney.

Only a few cases of lymphosarcoma of the tonsil in children have been reported. In two of these in which mention was made of roentgen therapy, it was without effect.

Prevention of Recurrence of Nasal Polyps. A Comparison of Results of the Postoperative Use of Radium and Zinc Ion Transfer. A. R. Hollender. *Ann. Otol., Rhin. & Laryng.* 56: 932-936, December 1947.

The extirpation of nasal polyps is best performed by classical surgical technics. For the prevention of their recurrence radium therapy has definite advantages. The author has used the method of Scal (*Arch. Otolaryng.* 16: 199, 1932), applying a 50-mg. capsule of radium, screened by a platinum container encased in gutta-percha, against the operated area in the nasal chamber for three hours. As a general rule, two to three treatments are given at intervals of two to three weeks. With this treatment, the regrowth of nasal polyps may be prevented in approximately 70 per cent of patients. The treatment, however, is expensive, radium is not always readily available, and the rhinologist with little experience in its application may hesitate to use it.

The author has found zinc iontophoresis, which he calls zinc ion transfer, an equally effective method. After surgical extirpation of all accessible polypoid tissue, two weeks are allowed for healing. Both nasal chambers are then treated at the same sitting. The treatment should be repeated in two to three weeks. Rarely is a third treatment necessary.

In 65 patients treated postoperatively with radium, the recurrence of nasal polyps was completely prevented in 47 and partially prevented in 18. In 72 patients treated postoperatively with zinc ion transfer, recurrence of polyps was definitely prevented in 55 and partially in 10. In 7 the treatment failed to produce the desired effect.

The claims of certain authors that zinc ion transfer produces severe reactions, anosmia, and other harmful effects have not been borne out in several thousands of treatments.

STEPHEN N. TAGER, M.D.

Treatment of Felons with X-Rays. P. Funck-Brentano and J. H. Marchand. *Presse méd.*, July 5, 1947. (*Abst. in Prensa méd. argent.* 35: 62, Jan. 2, 1948.)

The action of the x-rays in acute infections, though not well established, is assumed to be an excitation of

the cellular defenses, the stimulation of the reticulo-endothelial tissues, vasodilatation, and destruction of the leukocytes, with resultant liberation of antitoxins. Experience teaches that the more acute the infection, the smaller should be the dose of x-rays. Two or three sessions of 50 to 100 r are sufficient for a florid lesion; a subacute or chronic lesion may require 10 or 12 applications of 100 to 200 r.

The following factors are employed: 100 to 150 kv.; 5 mm. Al or 0.5 mm. Cu plus 2 mm. Al; target-skin distance 25 to 30 cm. The field of irradiation covers the inflammatory zone plus a margin of 1 or 2 cm. In case of felon with osseous involvement, and particularly osteitis, radiotherapy produces prompt relief of pain. Where surgery cannot be avoided, it is practised with greater security, as the danger of diffusion of the infection is minimized.

JAMES T. CASE, M.D.

Technic and Problems of Roentgen Ray Epilation.

Frank C. Combes and Howard T. Behrman. Arch. Dermat. & Syph. 57: 74-85, January 1948.

The authors use the five-point technic of Kienböck and Adamson for roentgen-ray epilation in cases of epidemic ringworm of the scalp. They differ from many authorities in the daily use, both before and after epilation, of tincture of green soap followed by the application of ointments containing 5 and 10 per cent sulfur or ammoniated mercury. The radiation factors preferred are 100 kv., 5 ma., 20 cm. distance, h.v.l. 1.0 mm. Al. No filtration is used other than that inherent in the tube (0.5 mm. Al), and the rate of exposure is 100 r per 30 seconds, 350 r being delivered in one

minute and forty-five seconds. No cases of permanent alopecia have been observed among hundreds of cases, and it is believed that the margin of safety is probably much greater than it is generally assumed to be. Full details of preparation of the patient and of the irradiation technic are included.

Histoplasmosis. Cutaneous and Mucomembranous Lesions, Mycologic and Pathologic Observations. Hiram E. Miller, Frances M. Keddie, Herbert G. Johnstone, and Warren L. Bostick. Arch. Dermat. & Syph. 56: 715-737, December 1947.

A study of 88 cases of histoplasmosis recorded in the literature reveals that cutaneous or mucomembranous lesions were present in 45. *Histoplasma capsulatum* was observed in a large percentage of these cutaneous lesions.

An additional case of histoplasmosis is reported in a man in whom an ulcer of the tongue developed. The diagnosis was established five months before death on isolation of the fungus in cultures of biopsy tissue from the walls of the ulcer. In addition to the involvement of the tongue, the patient had histoplasmosis of the cervical and axillary lymph nodes with a possible terminal invasion of the blood stream by the parasite. He had an associated Hodgkin's disease of five years duration and disseminated miliary tuberculosis, which probably caused his death. Treatment with sulfadiazine, stilbamme glucoside, ("neostam") and penicillin was of no value. Roentgen therapy (1,000 r) was given to the left half of the ulcer; no appreciable effect was noted. The mycologic, histologic, and postmortem observations are described in detail.

RADIOACTIVE ISOTOPES

Use of Radioactive Isotopes in Diagnosis and Therapy. J. R. Maxfield, Jr., and Jack G. S. Maxfield. Texas State J. Med. 43: 558-561, January 1948.

This paper is in the nature of a summing up of the present status of radioactive isotopes in diagnosis and therapy, with occasional comments on the authors' personal experience. Since they emit ionizing radiations, radioactive isotopes may be used in minute quantities as tracers and in larger amounts therapeutically for their biological effect. While some 400 of these isotopes are possible of production, only a few have been employed medically. By far the most commonly used are radioiodine and radiophosphorus.

Among the diagnostic applications, the authors mention (1) the use of radioactive iodine (a) as an adjunct in the diagnosis of early hyperthyroidism and myxedema, (b) in assessing the effectiveness of thiouracil therapy, and (c) in tracing distant metastases in selected cases of carcinoma of the thyroid; (2) the use of radiophosphorus, as suggested by Low-Beer (Radiology 47: 492, 1946) for predicting the nature of masses in the breast and axilla and for the determination of the radio-sensitivity of enlarged lymph nodes, especially of the lymphoblastoma group; (3) the use of radioactive sodium and radioactive phosphorus in determining the adequacy of the circulation in the presence of vascular disease, thus indicating where amputation can be carried out at the most distant point for vitalization of tissue and with minimum sacrifice of an extremity.

In the field of therapy, there are two conditions which stand out above all others as ideal for the use of radio-

active materials: polycythemia vera and hyperthyroidism or thyrotoxicosis. Radioactive phosphorus is the present treatment of choice in the former condition, bringing about prolonged remissions in some 80 per cent of the cases. In thyrotoxicosis, also, it has produced good results in a high proportion of cases, without surgical removal of the thyroid and without danger of serious complications.

Desirable palliative effects have been obtained in chronic leukemia by the use of radioactive phosphorus, and the authors believe there is a selective increase in the duration of life over other methods of treatment, though this is unproved. The disagreeable effects of radiation sickness and of the skin reactions sometimes inevitable with doses of roentgen therapy sufficient to produce regressions are avoided, and cases that have become refractive to roentgen rays may sometimes respond to isotope therapy. In certain instances where there is a markedly enlarged liver or lymphadenopathy producing symptoms of obstruction or severe discomfort, the authors have found localized roentgen irradiation directed to these areas plus radioactive phosphorus in systemic distribution a valuable method of treatment. In the acute leukemias, radioactive phosphorus, like other methods of therapy, has little to offer. It may, however, produce brief remissions.

Although the number of thyroid carcinomas that will absorb iodine is small, treatment by radioiodine is still worthy of consideration. The authors have found that low-grade metastasizing carcinomas with an increased basal metabolic rate and some evidence of thyrotoxic-

is are most likely to respond favorably. They point out the necessity of individualized treatment and the futility of attempting to lay down hard and fast rules.

The use of various radioactive colloids in neoplastic disease is being pursued at various centers. Mention is made of insoluble chromic phosphate for lesions of the liver and spleen and of the pleura and peritoneum, and of radioactive gold colloid and radioactive manganese dioxide colloid in lymphoblastoma. Chromium phosphate has been injected beneath malignant tumors of the skin, and basal-cell lesions and keratotic lesions have been treated by application of radioactive materials by the blotting paper technic of Low-Beer (*Radiology* 47: 213, 1946).

In conclusion, the authors warn that isotopes should be used only by personnel competent to handle and administer such materials. They place upon the radiologist the responsibility of calling to the attention of his colleagues in other branches of medicine the hazards involved in the use of artificial radioactivity and of teaching all those in contact with the project correct methods of handling these materials.

ERNEST S. KEREKES, M.D.

Use of Radioactive Iodine in the Study of Thyroid Function in Man. E. B. Astwood and Malcolm M. Stanley. *West. J. Surg.* 55: 625-639, December 1947.

Sixty-six normal subjects and 20 patients with disordered thyroid function were studied in an attempt to develop a practical method for the diagnosis of thyroid disease and the evaluation of antithyroid compounds. The material used was ¹³¹I without added carrier iodine, given orally in sodium chloride solution. A dosage of 0.1 millicurie was found to be satisfactory in most instances. Measurements of radioactivity were made with a shielded Geiger counter centered over the anterior aspect of the neck.

During the early hours of iodine uptake by the thyroid gland, the curve of increasing concentration was found to approximate a parabola. The slope of the straight line obtained by plotting the increasing radioactivity against the square root of the time in minutes provided an arbitrary numerical expression of the rate of uptake and was referred to as the accumulation gradient. This method of plotting permitted a prediction of the expected course of uptake when frequent measurements were made during the first two to three hours, and it facilitated a study of the influence of iodine, thiocyanate, and antithyroid compounds.

The rate at which the thyroid accumulated iodine varied widely in normal individuals; the gradients averaged 9.3 but varied between the limits 1.5 and 36.4. Four myxedema patients had very low gradients, as did four who received ordinary iodine. Hyperthyroidism was associated with concentration gradients greater than the normal average but only one of seven hyperthyroid patients had a gradient beyond the normal range. A single dose of 100 mg. of propylthiouracil

completely stopped the accumulation of iodine within thirty minutes, and the effect lasted for three hours; larger doses gave a more prolonged inhibition. Propylthiouracil given before the radioactive iodine markedly limited the uptake of radioactive iodine; the small amount of iodine accumulated under these conditions remained as iodide and was discharged by giving potassium thiocyanate and diluted out by the administration of potassium iodide. MAURICE D. SACHS, M.D.

Treatment of Graves' Disease with Radioactive Iodine. Earle M. Chapman. *West. J. Surg.* 56: 47-51, January 1948.

As a result of experience with radioactive iodine (¹³⁰I) with a half-life of twelve hours in the treatment of 45 patients with diffuse goiters or hyperthyroidism, the author concludes that twelve-hour iodine is an effective single therapeutic agent. Of the 45 patients treated, 33 responded to a single dose, 3 to two doses, 5 to three doses, and 4 failed to respond. Contraindications to use of the isotope are pregnancy and renal disease.

MAURICE D. SACHS, M.D.

Use of Radioactive Sodium as a Guide to the Efficacy of Drugs Used in Treatment of Diseases of the Peripheral Vascular System. Preliminary Report. Isidor Mufson, Edith H. Quimby, and Beverly C. Smith. *Am. J. Med.* 4: 73-82, January 1948.

Objectives in the treatment of peripheral vascular diseases are the reopening of completely or partially closed blood vessels and the development of an adequate circulation. The vasodilator drugs, papaverine hydrochloride and histamine, have frequently been reported as useful in obtaining such results. A preliminary report is made of the use of radioactive sodium as a guide to the efficacy of these drugs. The authors' conclusions are as follows:

1. Patients with scleroderma, thrombo-angiitis obliterans, obliterative arteriosclerotic endarteritis and non-specific arteritis of the minute vessels show a sub-normal curve for the diffusion of radioactive sodium from the blood vessels.
2. Histamine administered by iontophoresis or by intra-arterial injection brought about a definite rise in the diffusion curve. When given intravenously in one patient it produced the opposite effect.
3. The dilatation caused by histamine was not reversed by 1:250,000 adrenalin given intravenously.
4. Neither papaverine nor 5 per cent sodium chloride solution given intravenously produced changes in the radioactive sodium curves.
5. Of the drugs tested, only histamine appeared able to produce an immediate increase in the capillary diffusion rate as determined by a radiosodium curve taken following a single dose.
6. A method for giving an intra-arterial infusion is described. To date ninety such infusions have been administered.

EFFECTS OF RADIATION

Radiation Thrombocytopenia (Preliminary Report). Harry Mossberg. *Acta radiol.* 28: 110-114, April 30, 1947.

A thrombocytopenia and a hemorrhagic diathesis following teleroentgen therapy and after experimental ir-

radiation of animals have been reported by several investigators. Except for two previously published cases of his own (*Acta radiol.* 27: 400, 1946), however, in which a fatal hemorrhagic diathesis occurred, the author was unable to find any report in the literature of a simi-

lar condition appearing after "ordinary" local roentgen treatment to relatively small fields in patients with relatively normal blood cell counts.

The investigation reported here had to do with the effect of radiation on the thrombocytes and blood clotting in 40 patients. A definite reduction in the number of platelets was found in two-thirds of the cases; in some instances this occurred during the last week of treatment or immediately after treatment, and in others four to six weeks later. In some cases the late thrombocytopenia was more pronounced than the initial reduction. Five patients, in addition to the thrombocytopenia, had clinical signs of hemorrhagic diathesis; in 2 of these it was considered the cause of death. In one patient who had received relatively small roentgen doses over the entire vertebral column the bleeding time was increased to an hour.

Radiation over the thorax, sternum, and vertebral column is most apt to produce thrombocytopenia, and the author believes that it is very important to follow the thrombocyte curve in these cases, especially at the end of the treatment series and during the following five or six weeks.

Physiology and General Management of Chronic Ulceration Occurring After Irradiation. Howard B. Hunt and Donald H. Breit. *Am. J. Roentgenol.* 59: 9-17, January 1948.

Persistent or recurrent ulceration following irradiation is influenced by a number of factors, namely: (a) constitutional disease (arteriosclerosis, deficiency states, syphilis, etc.), (b) impaired blood supply, (c) infection, (d) presence of devitalized tissue, (e) trauma of any type, (f) inadequate regeneration from any cause, and (g) residual or recurrent tumor.

Each of these factors is discussed fully and various means of prevention and treatment are given. Animal studies show that greasy ointments increase the severity of erythematous reactions while water or dry powder does not.

Radon ointment definitely stimulates the regeneration of epithelium, but the defects should be kept at a minimum. Surgical aspects are to be presented more fully in a subsequent paper.

ZAC F. ENDRESS, M.D.

Spindle Cell Epidermoid Epithelioma Simulating Sarcoma in Chronic Radiodermatitis. Charles F. Sims and Neville Kirsch. *Arch. Dermat. & Syph.* 57: 63-68, January 1948.

In 2 cases of chronic radiodermatitis of the face, of many years duration, a neoplasm developed. In both instances, the growth closely resembled sarcoma histologically, but closer study with special staining showed spindle-cell epidermoid epithelioma. The spindle-shaped cells were derived from the prickle-cell layer of the epidermis, and their appearance had changed so that they took on an elongated narrow fusiform appearance, resembling spindle-shaped connective-tissue cells. These morphologic changes may be due to the associated sclerosis and fibrosis present in the irradiated skin, the squeezing and compression effects altering the cell shapes. True sarcomas developing in a roentgen or

radium dermatitis are a rarity and have yet to be proved.

Obstructive Lesions of the Small Intestine and Sigmoid Due to Irradiation. James C. Spackman. *Ann. Surg.* 127: 121-127, January 1948.

The author points out that a patient who has received irradiation to the pelvic region and later complains of abdominal pain, diarrhea, passage of mucus or blood, or vomiting, does not necessarily have a recurrence or extension of the original disease. Such symptoms may be due to a partially obstructing lesion, the early or late result of radiation therapy. This lesion may be in the terminal ileum or lower sigmoid. Since the terminal ileum is frequently in the low pelvis, symptoms of complete or incomplete low small bowel obstruction often appear earlier than evidence of obstruction of the sigmoid. The early symptoms are caused by hyperemia and mucosal edema of the segment of bowel involved, and may occur as early as two weeks following irradiation. Later complications (perhaps after several years) are due to fibrosis and cicatricial contraction.

Abdominal exploration after appropriate study and observation is indicated. The obstructing lesions may be corrected by proper surgery.

Five case reports are presented. Three of the patients had received radiation therapy for carcinoma of the cervix, one for papillary cystadenocarcinoma of the ovary, and another for carcinoma of the corpus uteri.

Excellent roentgenograms from two of the cases are reproduced.

EDWARD E. LEVINE, M.D.

Comparison of the Lethal Effect of Neutrons and Gamma Rays on Mouse Tumours (a) by Irradiation of Grafted Tumours *in vivo*, (b) by Irradiation of Tumour Fragments *in vitro*. L. H. Gray and John Read. *Brit. J. Radiol.* 21: 5-10, January 1948.

Studies of the effects of neutrons and gamma rays on mouse carcinoma 2146 *in vivo* and *in vitro* were made. Discounting a probable error of 20 per cent in measurement of the neutron dose and 3 per cent in the gamma ray dose, the neutron irradiation was found to be twenty-four times more effective *in vivo* and 9.5 times more effective *in vitro*. This is considerably higher than has been found by other workers. Comparison with previous reports indicates that the efficiency of neutron irradiation decreases with increasing energy.

SYDNEY J. HAWLEY, M.D.

Further Development of the Theory of Biological Reactions Caused by Roentgen Rays. J. Th. van der Werff. *Acta radiol.* 28: 169-184, April 30, 1947.

The author further amplifies his theory of the mathematical relationships of the biologic effects of roentgen rays, which were published originally in 1942 (*Acta radiol.* 23: 603, 1942. *Abst. in Radiology* 45: 215, 1945). The equations as developed correlate well with the experimental results obtained by Forssberg on the fungus *Phycomyces blakesleeana*. A monograph giving the relationship to the "hit-theories" and the results of dosage distribution is planned.

ELIZABETH A. CLARK, M.D.

November 1944

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